

NEWBORN SCREENING IN NEBRASKA

Newborn Bloodspot Screening for
Metabolic and Inherited Disorders

and

Early Hearing Detection & Intervention



2016
Annual Report

NEBRASKA

Good Life. Great Mission.

DEPT. OF HEALTH AND HUMAN SERVICES

2016 Annual Report Newborn Bloodspot Screening

TABLE OF CONTENTS

NEWBORN SCREENING FOR INBORN ERRORS OF METABOLISM AND INHERITED DISORDERS	3
What it is and Why we do it	3
Newborns Identified in 2016	4
System Overview	5
MAJOR INITIATIVES OF 2016	
Educational Initiatives	6
Quality Assurance and Improvement Initiatives	6
Nebraska Ranking for Timeliness	7
Consideration of New Conditions	8
HIGHLIGHTS FROM SOME KEY ELEMENTS OF NEWBORN SCREENING	
Follow-up	9
Advisory Committee	10
Financing Newborn Screening	11
Assurance of Treatment with Metabolic Foods & Formula	11
Intervention Data	12
Outcome Data	13
STAFF	21
DISCLOSURES	22

NEBRASKA EARLY HEARING DETECTION AND INTERVENTION
2016 ANNUAL REPORT
TABLE OF CONTENTS

INTRODUCTION	14
NEWBORN HEARING SCREENING DATA REPORTED FOR 2016	15
Birthing Facility Screening Programs	15
Hearing Screening at Birthing Facilities and Birthing Centers	16
Parent Education	15
Monitoring, Intervention, and Follow-up Care	15
Testing Status of 2016 births	16
Lost to Follow Up Progress over 10 years	16
Timeliness of Follow-up Screening/Evaluations/EDN Services	17
ACTIVITIES – 2016	17
Funding	17
Advisory Committee	17
PROJECTS	18
HearU Nebraska/Children’s Hearing Aid Loaner Bank	18
Task Force	18
Hospital Site Visits	19
SUMMARY	19
STAFF	21
DISCLOSURES	22

NEWBORN SCREENING FOR INBORN ERRORS OF METABOLISM AND INHERITED DISORDERS



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. Problems with physical growth, 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, laboratory, 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.
- ❖ Assuring 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 2016 this resulted in identifying and treating 57 newborns in time to prevent or reduce problems associated with identified conditions.

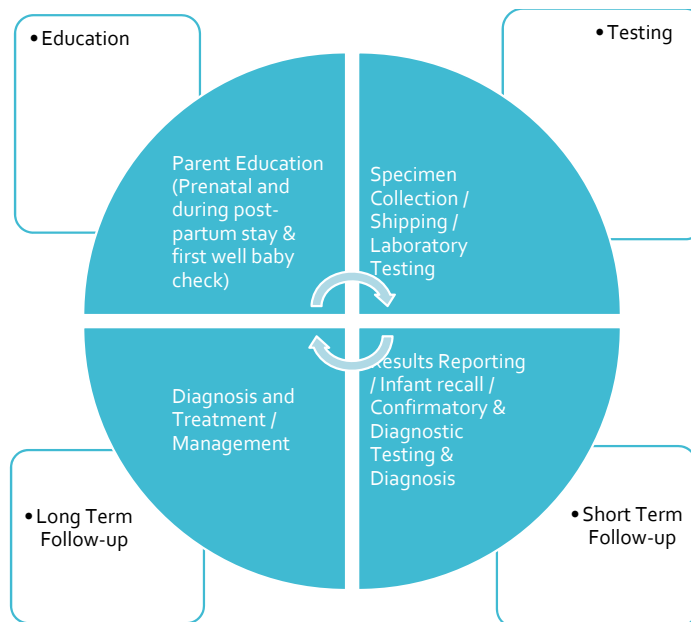
- 2 babies with Partial Biotinidase Deficiency (BIO)
- 1 baby with Cobalamin C Disease
- 1 baby with Compensated Hypothyroidism
- 2 babies with Congenital Adrenal Hyperplasia
- 8 babies with Congenital Primary Hypothyroidism (CPH)
- 7 babies with Congenital Hypothyroidism
- 1 baby with Hyperphenylalaninemia
- 1 baby with Hypothyroidism
- 6 babies with Cystic Fibrosis (CF) (1 case mild CF)
- 2 babies with CF Related Metabolic Syndrome (CRMS)
- 1 baby with Galactosemia
- 1 baby with Medium Chain Acyl Co-A Dehydrogenase Deficiency (MCAD)*
- 2 babies with Phenylketonuria (classical PKU)
- 2 babies with Primary Hypothyroidism
- 2 babies with Sickle Cell Disease
- 5 babies with Sickle Hemoglobin C Disease
- 1 baby with Sickle Cell Beta Thalassemia
- 1 baby with Sickle Beta Thalassemia + possible Alpha Thalassemia
- 1 baby with Beta Thalassemia Major
- 1 baby with Hemoglobin E Beta O Thalassemia
- 1 baby with Hemoglobin H Disease
- 1 baby with Transient Hypermethioninemia (not treated)
- 2 babies with Transient Tyrosinemia (1 treated)
- 2 babies with Very long-chain Acyl- CoA Dehydrogenase Deficiency (VLCAD)
- 1 baby with partial DiGeorge Syndrome 22q11.2 deletion (from SCID screen)
- 1 baby with idiopathic T-Cell lymphopenia (from SCID screen)
- 1 baby with 3-methylcrotonyl coxylase deficiency (3-MCC)

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



System Overview

In 2016, fifty five birthing facilities in Nebraska shipped specimens overnight for 27,036 babies Monday through Saturday to PerkinElmer Screening Laboratory (all but 3 hospitals have Saturday pick up available to them). The program, administered by the NDHHS, 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, Nebraska Chapter of the March of Dimes, 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 assure 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 2016

❖ EDUCATIONAL INITIATIVES

- Two Grand Rounds presentations were given by NDHHS staff at Children's Hospital in May. "Newborn Screening 101" and "Special Considerations in Newborn Screening...Perils, Pitfalls, and Pearls of Wisdom" were well received.
- A "Practitioner's Update" was sent to all Family Physicians and Pediatric practices in Nebraska addressing the Practitioner's responsibilities for newborn screening education, ordering tests, following up, and referral and treatment. Clinic practices were encouraged to triage incoming faxes for quicker response time.
- Timeliness! That was the major theme of the year, always looking for ways to improve. One effort included providing laminated posters to hospital laboratory and mother/baby units emphasizing the steps in the process and reminding personnel that "Every Hour Counts!"

❖ QUALITY ASSURANCE AND IMPROVEMENT INITIATIVES

The program 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.
- In 2016 Nebraska's Newborn Screening Program was awarded a "Timeliness" grant from the NewSTEPS program of the Association of Public Health Laboratories (APHL) and the Colorado School of Public Health. In 2016 the program awarded \$10,000 contracts to 4 hospitals to help them achieve bi-directional electronic interfaces with the newborn screening laboratory. This 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 a quarterly report 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.
 - Quarterly reports in the first half of the year were accompanied with a count of the number of incidents of batching and late specimen collection in the last quarter, as well as the number of incidents of any babies who were discharged without a screen in the prior year.

- During the second half of 2016 the weekly batching and late specimen collection monitoring had to be suspended due to increased demands on existing personnel.
- 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 5 days of age, and the percent of all results reported out by 7 days of age.
- The US Government Accountability Office released a report to Congressional Committees on Newborn Screening Timeliness in December of 2016. Although the publication doesn’t identify states by name, the data that was submitted by NewSTEPS 360 and used for this report showed Nebraska ranked favorably in comparison to other reporting states on several measures:

Nebraska Ranking as reported by NewSTEPS 360

Year	Specimen collection within 48 hours of birth	Rank	Specimen Receipt within 48 hours of Collection	Rank
2012	98.8%	2 of 33	53.3%	10 of 32
2013	98.9%	2 of 33	53.2%	10 of 32
2014	98.9%	3 of 35	54.3%	11 of 33
2015	99.3%	2 of 35	58.2%	14 of 34

Year	Reporting time-critical results* within 5 days of birth	Rank	Reporting all results within 7 days of birth	Rank
2012	40%	5 of 14	97.8%	1 of 27
2013	71.4%	3 of 14	98.1%	1 of 27
2014	37.5%	6 of 15	97.9%	2 of 28
2015	0%	5 of 16	98.1%	2 of 27

*The Advisory Committee on Heritable Diseases in Newborns and Children has defined the “Time Critical” diseases as: PA, MMA, IVA, 3-HMG, MCAD, VLCAD, LCHAD, TFP, ASA, CIT I, MSUD, GAL, CAH, MCD, BKT and GAI.

In addition at the 2016 Timeliness Grantee Meeting Nebraska received the following NewSTEPS 360 timeliness state ranking data:

Figure 5: Specimen Collection within 48 Hours of Birth

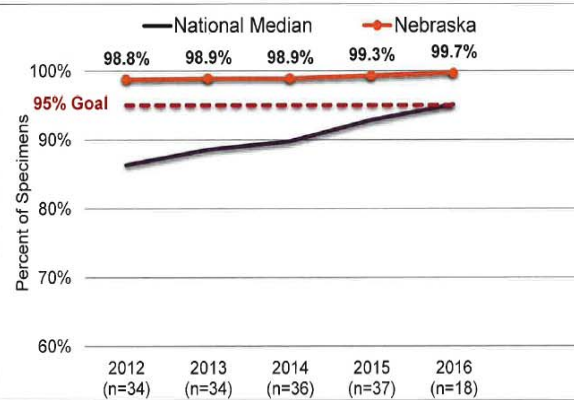


Figure 6: 2015 State Rank for Specimen Collection within 48 Hours of Birth (n=37)

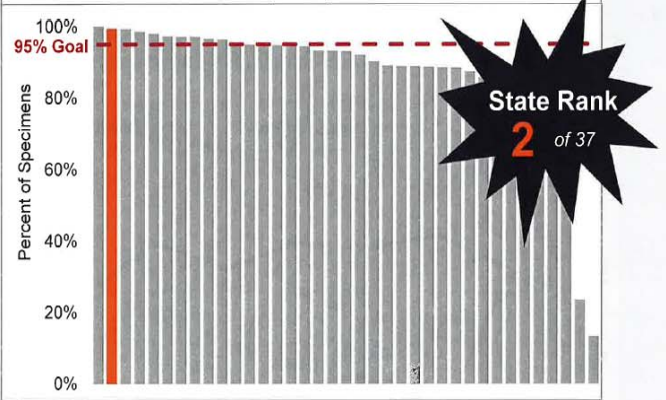


Figure 3: Reporting All Results within 7 Days of Birth

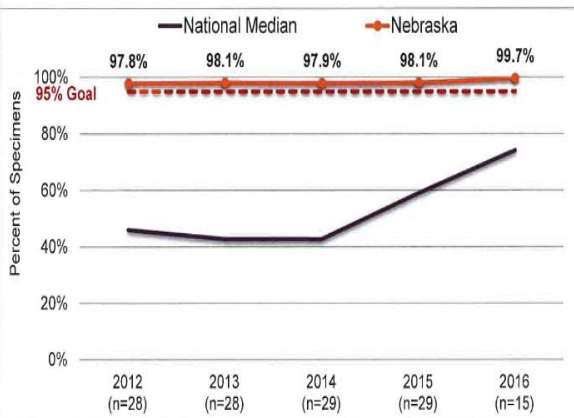
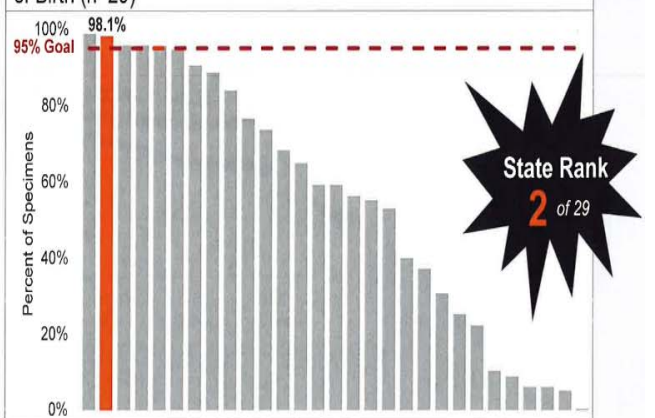


Figure 4: 2015 State Rank for Reporting All Results within 7 Days of Birth (n=29)



- Three new conditions were recommended for addition to the Newborn Screening Panel by Nebraska’s NBS Advisory Committee. The Program applied for and was awarded an implementation grant (funding available till August 2018) to prepare for adding these new conditions to the screening panel. This funding will help support a part time follow-up, part time quality assurance staff person. Adding this capacity will allow 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 should 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) 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 2016 alone this was an essential function for more than 4000 results or distinct reasons for follow-up.

Many things are known to complicate and delay follow-up. An example of this is if the baby’s doctor identified on the filter paper (and to the follow-up program) is incorrect, and difficulties arise in locating the correct provider. Other things complicate follow-up such that any one baby can have multiple reasons for needing additional testing. Babies admitted to the neonatal intensive care unit (approximately 9% of all births) are especially prone to having complications with their newborn screen. For example, a baby can have an initial unsatisfactory specimen followed by a repeat specimen collected too close to the time of the last transfusion, followed by a result of multiple amino acid elevations consistent with hyperalimentation, or an inconclusive result for cystic fibrosis, all of which require monitoring and follow-up to ensure that an acceptable screen for all diseases is completed.

The two professional follow-up coordinators are highly collaborative. In today’s world with multiple baby name changes, and baby’s physician changing due to parent choice or insurance coverage changes, the follow-up coordinators 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 2016 the Committee recommended adding three conditions to the required newborn screening panel: Pompe, MPS-I and X-ALD.

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 (St. Paul) for chairing the Committee along with Dr. Samuel Pirruccello (UNMC) who served as Vice Chair from 2014 through 2016.

2016 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 Advisor
Sara Frink	Parent 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 Advisor
Rose Kreikemeier, APRN	Children's Hospital / Metabolic Management Clinic
Mary Larsen	March of Dimes, Nebraska Chapter
Richard Lutz, MD	Nebraska Medicine / Children's Metabolism/Genetics
Bev Morton	Parent 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
Ebrahim Shakir, MD	Midwest Allergy and Asthma Clinic, PC / Immunology
Jill Skrabal, MS, RD, LMNT, CDE	Metabolic Management Clinic UNMC
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).

Assurance of Treatment and Management of Inborn Errors in Metabolism



To fulfill the statutorily-required public health assurance role, NDHHS contracted with the metabolic clinic through the University of Nebraska Medical Center to provide nutritional counseling and monitoring, and distribution of the metabolic formula. In 2016 the contract began requiring insurance to be billed first, before billing the cost of the formula to NDHHS. UNMC 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 and made a significant structural change in 2016 by moving to a reimbursement system instead of contracting with a vendor from which families could order foods. For individuals meeting financial hardship waiver criteria, the Department will purchase 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.

Jill Skrabal, RD, LMNT, CDE from the UNMC and Children's Hospital metabolic clinics coordinated a few family education day gatherings at local grocers in Kearney and Omaha. Assisted by Kathryn Heldt, RD (Children's Hospital) and Jessy Davis (NBS program), they helped families learn which modified foods meet the criteria for the pharmaceutically manufactured low protein foods 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 reporting out 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 resistance to confirmatory testing,
- problems locating parents because of inaccurate contact information.

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

In 2016 the average age of intervention for babies identified with each condition are listed below:

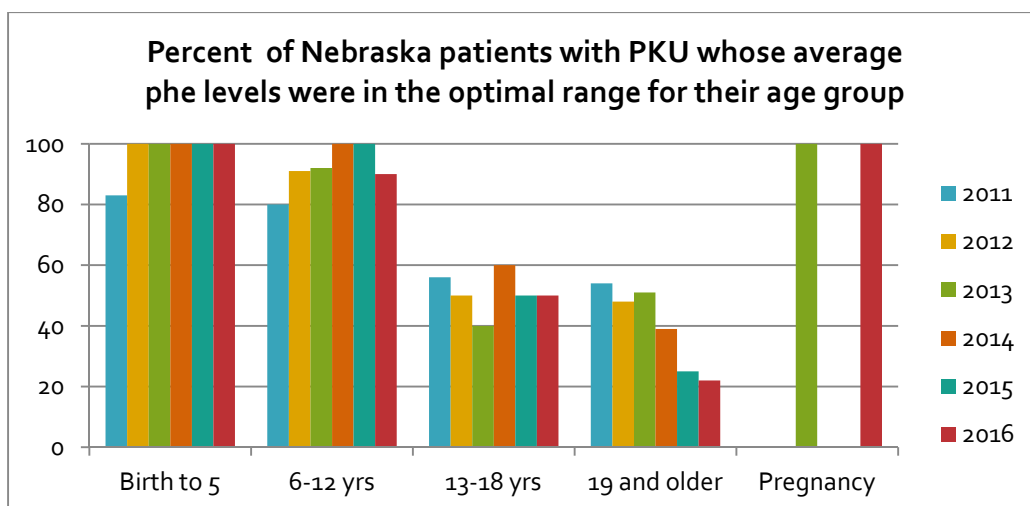
Partial Biotinidase Deficiency – 9 days
Congenital Adrenal Hyperplasia 26 days
Congenital Primary Hypothyroidism – 7 days
Congenital hypothyroidism – 46 days
Hypothyroidism – 10 days
Compensated hypothyroidism 83 days
Primary Hypothyroidism 39 days
Galactosemia – 4 days
Cystic Fibrosis - 16 days
CRMS - 79 days
MCAD - 5 days

Cobalamin C Deficiency -68 days
Sickle Cell Disease – 16 days
Sickle Beta Thalassemia – 14 days
Sickle Beta Thal + possible Alpha Thal – 51
Hemoglobin H Disease – 16 days
Hemoglobin E Disease Beta 0 Thal 130 days
Transient Tyrosinemia – 9 days
3-MCC Deficiency – 19 days
VLCAD – 4 days
Sickle Hemoglobin C Disease – 23 days
PKU 6.5 days



Outcome Data

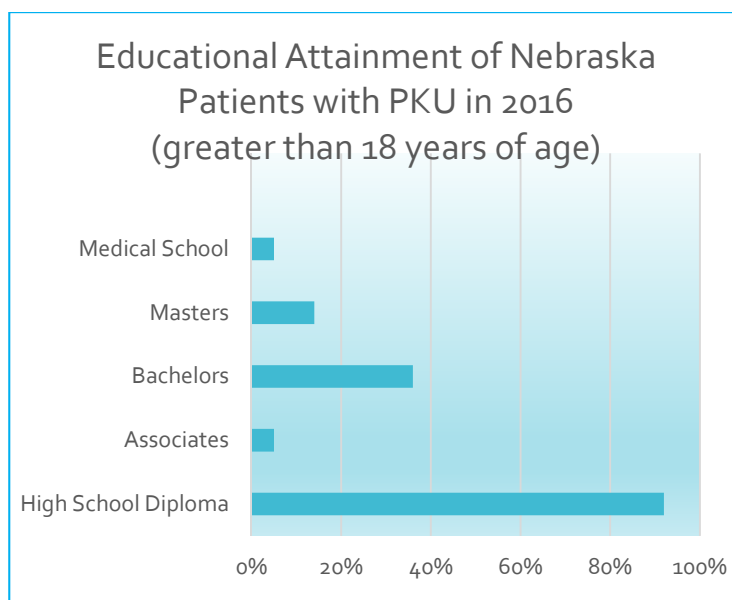
Measures to evaluate patient outcomes are important for evaluating the effectiveness of the newborn screening system. Close monitoring and ensuring access to the metabolic formula and foods are essential elements to the success of our Nebraskan's affected with inborn errors in metabolism.



Developmental Measures:

Since reporting of long-term outcomes began in 2011, through 2016, 100% of children with PKU age 0-12 receiving services in Nebraska were assessed by the pediatric metabolic specialists as meeting developmental milestones.

Outcome measures of educational achievement demonstrate the success of families, and individuals, made possible initially because of newborn screening:



NEBRASKA EARLY HEARING DETECTION AND INTERVENTION ANNUAL REPORT - 2016



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 (NDHHS) 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 2016 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 2016 there were 54 birthing facilities conducting hearing screenings.

Hearing Screening at Birthing Facilities and Birthing Centers



Photo courtesy of: NCHAM

In 2016, inpatient hearing screenings were reported on 26,878 newborns or 99.8% of the 26,925 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 2016, of those who received a screening in Nebraska, 25,967 (96.6%) passed the inpatient 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 2016. 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

NE-EHDI also provides a Parent Resource Guide to families with a child who has been diagnosed as deaf or hard of hearing to inform them about available resources. Feedback from

parents and the staff from the Guide By Your Side Program indicate the print version of the Parent Resource Guide is often overwhelming for parents to receive the day of diagnosis. Efforts have been made to make the guide more user friendly and easier to navigate, but the feedback remains the same. Therefore, an online version of the guide was added to the NE-EHDI website. This allows parents the option of receiving a print copy or accessing the information online. Use of the online version reduces the amount of time the NE-EHDI program staff uses to assemble the packets of information, and reduces funds spent on printing and shipping educational materials. The online version also allows information to be revised and updated as needed, instead of updates being available when materials are reprinted.



Photo courtesy of: Jordan Sochor Photography

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 2016, a total of 1,027 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 27,114 births for 2016:

26,810	Passed the screening or diagnostic testing (98.9% of births)
124	Expired (inpatient or outpatient)
42	Pending final screening or diagnostic testing
63	Diagnosed deaf or hard of hearing
37	Parents refused screening and/or diagnostic testing
22	Unresponsive (did not complete protocol after communication with NE-EHDI staff)
1	Lost (no response to NE-EHDI letters and phone calls)
11	Moved out of Nebraska
4	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.”

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 98% of infants received an inpatient screening within one month of age. For the newborns who were recommended for an audiologic diagnosis, nearly 68%

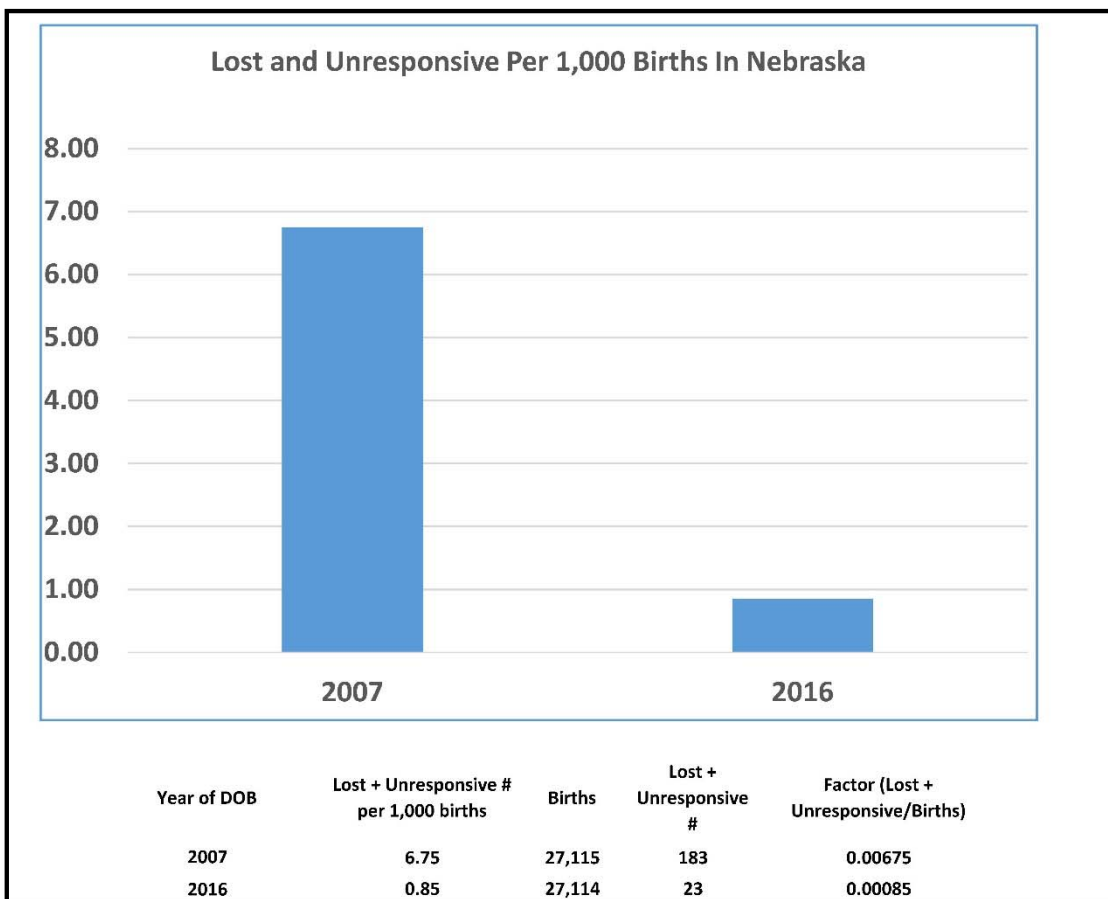
received the evaluation by 3 months of age according to individual data reported to the NE-EHDI Program by audiologists.

Note: Because 42 hearing records are still pending the final screening and diagnostic testing results, the “1-3-6” numbers above are preliminary as of June 2017 and will most likely change.

Records for the Early Development Network (EDN), Nebraska’s Part C Early Intervention Program, indicate that 76.5% of infants residing in Nebraska in 2016 diagnosed as deaf or hard of hearing with a developmental delay, were enrolled in EDN services by 6 months of age for families accepting Part-C services. The reasons for those infants not enrolled 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 10 years:

Nebraska Lost to Follow-up and Unresponsive (2007 vs 2016 DOB)



ACTIVITIES – 2016

Funding

The NE-EHDI Program continued to receive funding from the Health Resources Services Administration/Maternal and Child Health Bureau (HRSA/MCHB) and the Centers for Disease Control and Prevention (CDC). 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.

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



Advisory Committee meetings were held three times in 2016 and were open to the public.

Projects - 2016

HearU Nebraska

HearU Nebraska 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 2016. In 2016 there were 105 hearing aids provided and fitted for 57 children (age range of 1 month to 18 years) with “free” hearing aids provided by HearU Nebraska. Since 2008 over 317 children have been provided and fitted with hearing aids.

Task Force

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 during 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. Based on survey responses, the NE-EHDI Program will evaluate the quality of materials received by mothers in the hospital, the skills of the hospital employees responsible for conducting the hearing screenings, and the education parents receive about the results and next steps. The NE-EHDI Program will also survey hospital staff involved with the hearing screening process. Based on these survey results, NE-EHDI will develop additional training materials for staff who are responsible for educating parents about the results and next steps.

Hospital Site Visits



Photo courtesy of: Jordan Sochor Photography

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 lost to follow-up rates, 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.

Summary

- All of the 54 birthing hospitals in Nebraska conducted newborn hearing screenings in 2016 prior to discharge from the hospital or birthing center.
- In 2016, birthing hospitals reported screening the hearing of almost 99% of newborns prior to discharge from the hospital.
- Over 98% of the infants passed an inpatient screening, an outpatient screening, or a diagnostic evaluation.
- Of the 1,027 infants followed by the NE-EHDI Program, over 90% of these infants 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 2017.
- In 2016, there were confirmatory audiologic evaluations initiated within 3 months of age for 67% of newborns when the newborn did not pass the inpatient/outpatient screening or did not receive an inpatient screening.
- There were 113 infants born in 2016 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 2.3 per thousand screened is within the anticipated range of one to three per thousand.
- Nearly 77% of the infants identified 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.
- An online version of the guide was added to the NE-EHDI website. This allows parents the option of receiving a print copy or accessing the information online.
- The NE-EHDI website was nominated for Website of the Year at the EHDI 2016 Annual Meeting, and was among the top five EHDI websites in the nation.

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>

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:

Julie Luedtke, Newborn Screening/Genetics Program Manager 402-471-6733

Program planning, evaluation and management, professional and patient education, metabolic formula

Krystal Baumert, NBS Follow up Coordinator 402-471-0374

Metabolic and endocrine conditions, transfusions, home births

Karen Eveans, NBS Follow up Coordinator 402-471-6558

Hemoglobinopathies and cystic fibrosis

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>

E-mail contact: dhhs.newbornscreening@nebraska.gov

E-FAX: 402-742-2332

Regular Fax: 402-471-1863

Nebraska Newborn Screening Program
Department of Health and Human Services
P.O. Box 95026
Lincoln, NE 68509-5026

PerkinElmer Genetics Screening Laboratory Director, Joseph Quashnock, PhD 412-220-2300 (Pennsylvania)

PerkinElmer Genetics Screening, General Manager, PJ Borandi, 412-220-2300

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

Program planning, evaluation and management, systems development

Jim Beavers, Business Analyst, 402-471-1526

Data system planning and testing, development of reports, system security, training and technical assistance

MeLissa Butler, Community Health Educator Senior, 402-471-3579

Follow-up, patient education materials distribution, data management

Gabby Tachenko, 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, and education

Kiowa Rogers, Bilingual Community Outreach, 888-545-0935 (Contract, part-time)

Follow-up, community outreach, and education

Website: <http://dhhs.ne.gov/publichealth/EHDI/Pages/EHDIHome.aspx>

E-mail: DHHS.NEEHDI@nebraska.gov

Fax: 402-742-2395

Nebraska Early Hearing Detection & Intervention Program
Lifespan Health Services, Division of Public Health, DHHS
P.O. Box 95026
Lincoln, NE 68509-5026

The Nebraska Department of Health and Human Services is committed to affirmative action/equal employment opportunity and does not discriminate in delivering benefits or services.

This report was prepared and published by the Nebraska Department of Health and Human Services, Newborn Screening Program, P.O. Box 95026, Lincoln, NE 68509-5026.

*Filter Paper Blood spot photos courtesy of Whatman web site
[www.whatman.com/repository/documents/s7/51684%20\(S9036-812\).pdf](http://www.whatman.com/repository/documents/s7/51684%20(S9036-812).pdf).*

Laboratory photos courtesy of Perkin Elmer Genetics Screening Laboratory.

Any reference to specific commercial product in the Newborn Hearing Screening section does not constitute or imply an endorsement, recommendation or favoring by the Nebraska Early Hearing Detection & Intervention Program.

Newborn Bloodspot Screening is supported by the Health Resources and Services Administration (HRSA) of the U.S. Department of Health and Human Services (HHS) under grant number B04MC29351 Title V/Maternal & Child Health Services Block Grant Program for \$656,069.42 in Federal Fiscal Year 16, with zero percent financed with nongovernmental sources. This information or content and conclusions are those of the author and should not be construed as the official position or policy of, nor should any endorsements be inferred by HRSA, HHS or the U.S. Government.

The Nebraska Early Hearing Detection and Intervention Program is supported by the Health Resources and Services Administration (HRSA) of the U.S. Department of Health and Human Services (HHS) under grant number H61MC00065, Universal Newborn Hearing Screening, \$249,678 award amount for 4/1/2016 – 3/31/2017. No nongovernmental sources support the program. This information or content and conclusions are those of the author and should not be construed as the official position or policy of, nor should any endorsements be inferred by HRSA, HHS or the U.S. Government.

The Nebraska Early Hearing Detection and Intervention Program is also supported by the Centers for Disease Control and Prevention (CDC) under Cooperative Agreement Number 2NUR3DD000797-06-00, \$154,254 award amount for 7/1/2016 – 6/30/2017. The NE-EHDI Program is 100% financed with Federal money and no funding is received by nongovernmental sources. Its contents are solely the responsibility of the authors and do not necessarily represent the official views of the Centers for Disease Control and Prevention or the Department of Health and Human Services.