NEWBORN SCREENING IN NEBRASKA

Newborn Bloodspot Screening for Metabolic and Inherited Disorders

and

Early Hearing Detection & Intervention

2014 Annual Report

Department of Health & Human Services

DHHS

NEBRASKA

NEWBORN BLOODSPOT SCREENING
2014 ANNUAL REPORT
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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.

Newborn Screening is a system coordinated by the Nebraska Department of Health and Human Services Newborn Screening Program collaborating with hospitals, laboratory, health care professionals and families and involving 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.

In 2014, newborn screening for 29 conditions, resulted in successfully identifying and treating 60 newborns in Nebraska affected with conditions in time to prevent or reduce problems associated with them.

Newborns were diagnosed with these conditions from screening in 2014:

1 baby with Arginino Succinic Aciduria (ASA)
1 baby with Profound Biotinidase Deficiency (BIO)
5 babies with Partial Biotinidase Deficiency
7 babies with Congenital Primary Hypothyroidism (CPH)
9 babies with Congenital Hypothyroidism
3 babies with Hypothyroidism
1 baby with Primary Hypothyroidism (not congenital)
8 babies with Cystic Fibrosis (CF)
1 baby with CF Related Metabolic Syndrome (CRMS)
2 babies with Classic Galactosemia (GAL)
4 babies with Sickle Cell Disease
3 babies with Sickle Hemoglobin C Disease
2 babies with Sickle Cell Beta Thalassemia
1 baby with Hemoglobin E Beta Thalassemia
1 baby with Beta Thalassemia Major
1 Beta Thalassemia + Alpha Thalassemia Trait
1 baby with Sickle Hemoglobin C Disease plus Alpha Thalassemia Major
1 baby with Maple Syrup Urine Disease (MSUD)
1 baby with Medium Chain Acyl Co-A Dehydrogenase Deficiency (MCAD)
2 babies with Classical Phenylketonuria (PKU)
2 babies with Transient Tyrosinemia (1 treated/resolved)
1 baby with Hypertyrosinemia of Prematurity / Infancy
1 baby with Very Long Chain Acyl Co-A Dehydrogenase Deficiency (VLCAD)
1 baby with 3-Methyl Crotonyl Carboxylase Deficiency (3-MCC)

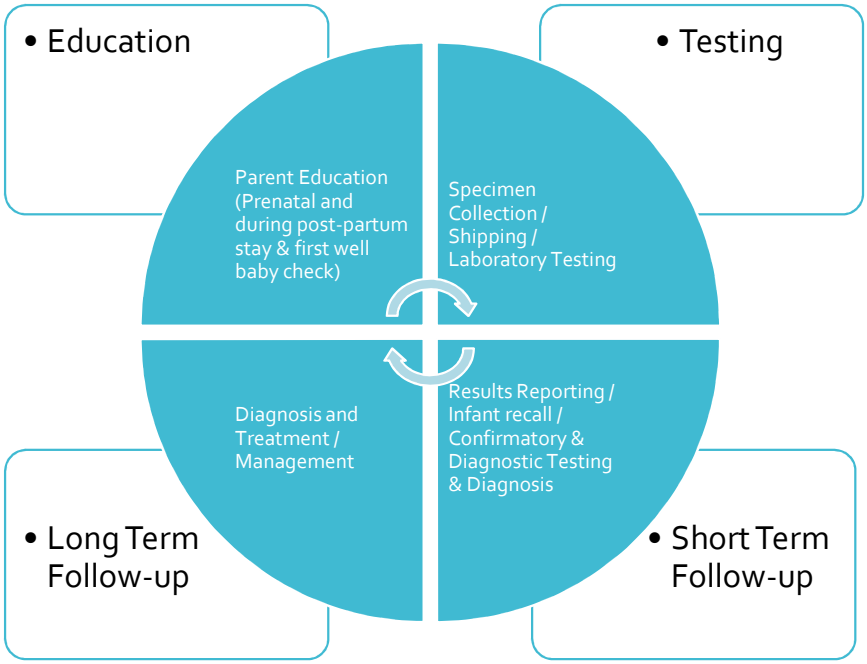
Individually each condition is rare, however the collective incidence of screened conditions in Nebraska 2006-2014 (424 diagnosed / 239,787 screened)

is

1 in every 565 births

System Overview

In 2014, 56 birthing facilities in Nebraska shipped specimens overnight, for 27,037 babies Monday through Saturday, to PerkinElmer Screening Laboratory. Perkin Elmer Genetics (Nebraska's contracted screening laboratory) expanded its agreement with UPS to ensure Saturday pick up at all Nebraska hospitals in 2014. The program, administered by the Department of Health and Human Services, 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 with experts from these subspecialties, laboratory, hospitals, pediatrics, neonatology, family practice, as well as 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 requires additional testing, received the necessary information to act accordingly. The program completed 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 a contract with a metabolic food vendor for individuals with PKU and other inborn errors in metabolism who require the specially manufactured foods. (For more information, visit our website at www.dhhs.ne.gov/nsp).



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

MAJOR INITIATIVES IN NEBRASKA DURING 2014

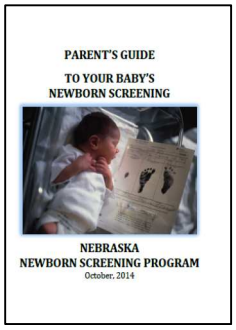
❖ SEVERE COMBINED IMMUNE DEFICIENCY SCREENING

Regulation revisions added screening for Severe Combined Immune Deficiency (SCID) effective in October 2014.

- To prepare, the SCID sub-committee/team (James L. Harper M.D., Russell Hopp D.O., Samuel Pirruccello M.D., and Ebrahim Shakir M.D.) with program personnel developed screening follow-up protocols for all newborns. Protocols were also developed for the population of newborns in the NICU expected to have higher rates of abnormal screens for SCID.

- The SCID team prepared practitioners for SCID screening via Pediatric Grand Rounds held at Children's Hospital in Omaha (with satellite feeds across the state) in June. The program sent educational updates to all birthing facilities as well as pediatricians and family physicians in Nebraska.

- The SCID Follow-up Coordinator Karen Eveans, MD also developed notification letters, Physician ACT (Action) sheets and parent information sheets used when notifying the baby's primary health care professional. Nebraska was about the 19th state to add SCID to the screening panel.

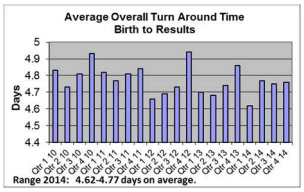


- Adding SCID to the screening panel provided the opportunity to update the "Parents Guide to Your Baby's Newborn Screening" with additional information about screening for SCID and screening for Critical Congenital Heart Disease (CCHD). The new information was provided in a one-page insert to hospitals until existing stock of the parent's guides were used. Distribution of the new parent's guide with the new information on SCID and CCHD began in December 2014 to the 56 birthing facilities, and upon request to some obstetric, family physician and pediatric practices. The Spanish translation was completed and is available as well.

- With the addition of SCID to the NBS Panel, the charge from the lab to the hospital, per infant screened, changed from $40.00 to $45.50.

❖ QUALITY ASSURANCE AND IMPROVEMENT INITIATIVES

The program continued its longstanding oversight and monitoring of multiple quality measures of hospital and laboratory performance while spearheading an initiative to revise and enhance the NBS QA system.



- The primary focus was on improving "TIMELINESS" which had become a national agenda item in the newborn screening field.

- An open invitation to hospital laboratory, nursery, NICU and QA leadership to participate in this revision resulted in 16 individuals representing 13 hospitals providing input on the new measures, benchmarks and reporting procedures.

- With the advice and input of the NBS Advisory Committee on the measures, benchmarks and report subsequently developed by the program, the laboratory worked during the last quarter of 2014 to develop the information technology (IT) capacity to produce this report quarterly.

- The new reports monitoring the time from birth to specimen collection, collection to receipt at the lab, in-lab processing time and birth to results time, now measure the *percent* of specimens from each hospital meeting various benchmarks. Previously the reports only measured the average turnaround times on each of those measures.

% meeting benchmark
vs.
Average performance on each measure

- A new procedure for reporting hospital violations of newborn screening regulations to the Acute Facility Licensing Unit in the Department of Health and Human Services was developed, and implemented.

- In addition to the weekly "batching/specimen shipping delay" report run by the program manager and reported to hospital QA contacts, a new report was developed in collaboration with PerkinElmer laboratory IT for monitoring initial specimens collected late at greater than 48 hours. This gives quicker feedback to hospitals and allows a more rapid response to systematically correct any problems.

- A poster on this new initiative was presented at the National Association of Public Health Laboratory's Newborn Screening Symposium held in Anaheim, CA.

❖ A COLLABORATIVE EDUCATION / QI INITIATIVE

The program reached out to nursing schools in 2014 to determine interest in receiving education about newborn screening or collaborating on newborn screening projects. The University Of Nebraska College Of Nursing Accelerated Nursing Program subsequently partnered with the program on a project in the Fall/Winter of 2014.

- The project was designed to determine Nebraska nurses' basic knowledge about newborn screening and opinions about their preparedness to educate others about it.

- Surveys went to all birthing facility nurseries and NICUs, family practice, pediatric and obstetric practices.

- Initial analysis of data by the program was discussed with the students, and presented to the NBS Advisory Committee. Further analysis will be conducted in 2015 to inform strategies for reaching out and providing more nurses with the knowledge and ability to be able to educate the patients and families with whom they work, about newborn screening.

- The nursing students were able to incorporate the project into their curriculum and report on the experience and what they learned to the rest of their class.

SURVEY RESULTS

107 of 585 nurses responded to the survey for an 18.2% response rate

A few key findings from the preliminary analysis of the responses found:

- 98% knew the blood sample was taken from the heel
- 63% knew 5 drops were taken
- 69% knew the specimen should be collected at 24-48 hours
- 71% knew the Affordable Care Act requires insurance to cover the newborn screening
- 56% felt somewhat or very comfortable educating parents about newborn screening
- 85% ranked the prenatal visit first, for appropriate times to educate new/expecting parents

HIGHLIGHTS FROM SOME KEY ELEMENTS OF NBS

❖ FOLLOW-UP

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 until either a satisfactory screen has been completed, an inconclusive result has had a repeat screen, or a positive result has received confirmatory testing and a diagnosis is either made or ruled out. In 2015 alone this was an essential function for more than 4200 results. Follow-up can be complicated and a single baby may need it for multiple reasons, for example: 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. The two professional follow-up coordinators are highly collaborative. In today's world with multiple baby name changes, and baby's physician's being changed 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 to followup. 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.

- Much of Nebraska's success can be directly tied to its recommendations and guidance! (Refer to the "contacts" page at www.dhhs.ne.gov/nsp for the list of fantastic advisors who provided technical expertise and policy guidance to the Nebraska Newborn Screening Program).

- Members committed at least a half day every three months to advise the state program (collectively over 250 hours). 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.

❖ 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).
The state general fund appropriation has stayed the same since 1997. Although the Title V Block Grant appropriation to the state is below 1997 levels, the proportion allocated to support Nebraska's Newborn Screening Program and state statutory requirements has increased substantially.

Assurance of Treatment And Management For Inborn Errors In Metabolism

To fulfill the statutorily-required public health assurance role, the Program contracted with the metabolic clinic through the University of Nebraska Medical Center to provide nutritional counseling and monitoring, and distribution of the metabolic formula.

The program coordinated the day-to-day pharmaceutically manufactured metabolic foods program by helping families understand the program and stay connected, monitoring vendors with which the department contracted and processing payments for family orders. Families received a tracking log for their use in monitoring their orders and expenses and they received an annual spending report. More intense work with the State's Department of Administrative Services on the re-bid for the commodity contracts resulted in a single vendor contract for foods. The NBS Program personnel adapted to a new required system of generating purchase orders and processing a three-way match in order to process payments to the vendors to meet new accountability and transparency expectations. During the last few months to ensure families could get the specialized foods that were otherwise unavailable, the program placed orders for families with another vendor that was carrying items not available from the contracted vendor. A plan to restructure the program to enhance family's buying power, increase the variety of foods they could access, and reduce the administrative burden on the program began to form in the latter months of 2014.

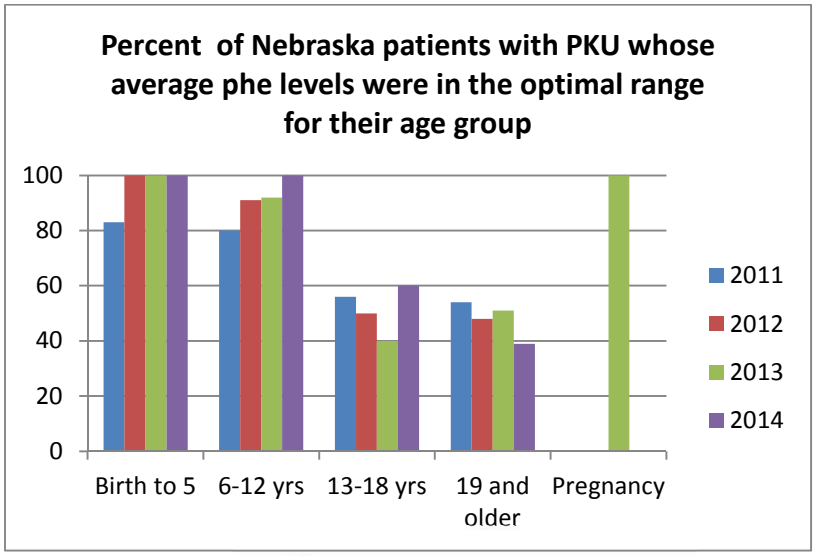
Intervention Data

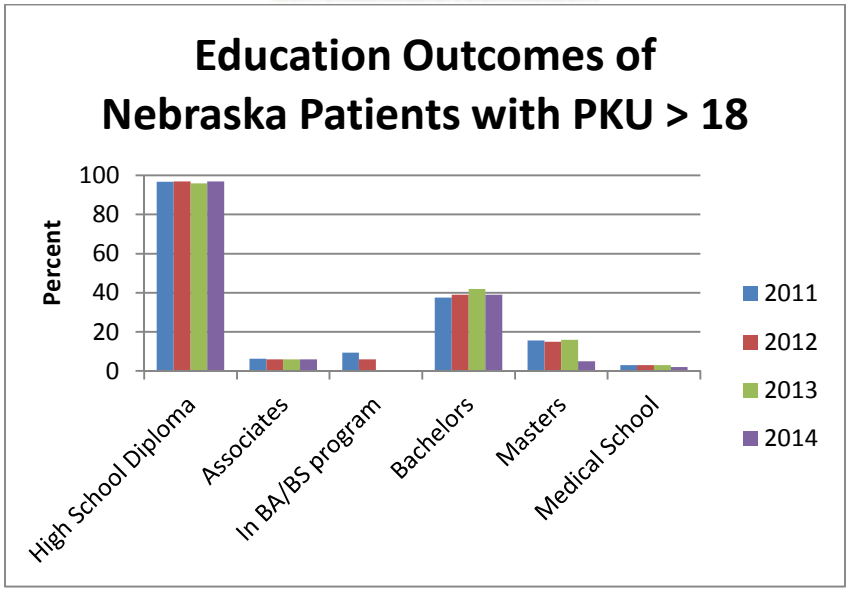
Collecting specimens correctly the first time, at the right time and processing them for shipment is just the beginning. Working to optimize shipping times with the commercial overnight shipping company, and maximizing the efficiencies at the testing laboratory are also key to reporting out results on 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 babies with prolonged treatment in NICUs, parental resistance to confirmatory testing, problems in locating parents because contact information provided to the hospital or recorded on the filter paper collection cards was incorrect or no longer accurate. All parts of the system must work to reap the best benefits of early identification, treatment and intervention.

Condition & number of babies diagnosed	Average age (days) at intervention/tx.	Range in ages (days) at intervention/tx.
1 Arginino Succinic Aciduria	4	4
1 Biotinidase Deficiency & 5 Partial Bio's (3 of the partial's treated)	12	7-17
7 Congenital Primary Hypothyroidism 9 Congenital Hypothyroidism 1 Primary Hypothyroidism 3 Hypothyroidism	22.2	CPH: 6-19 CH: 4-26 PH: 32 H: 6-14
8 Cystic Fibrosis and 1 CRMS	21.7	2-113
2 Classic Galactosemia	4	2-6
Hemoglobinopathies: 4 Sickle Cell Disease 3 Sickle Hgb. C Disease 1 Hgb. E Beta Thalassemia Disease 2 Sickle Beta Thalassemia 1 Beta Thalassemia 1 Hgb. SC Disease + Possible Alpha Thal 1 Beta Thalassemia + Alpha Thal Trait	24	Sickle Cell: 12-38 Sickle Hgb. C: 12-30 E Beta Thal: 18 S Beta Thal: 30-66 Beta Thal: 27 SC Dis + Alpha: 6
1 MCAD	6	6
1 MSUD	4	4
2 Transient Tyrosinemia (1 treated/resolved)	38	38
1 VLCAD (suspected)	6	6
1 3-MCC	32	32

Outcome Data

Measures to evaluate patient outcomes are important for evaluating the effectiveness of the newborn screening system. This data supports the need for continued funding for metabolic formula and foods required by patients identified with metabolic conditions via newborn screening. This data demonstrates it is money well spent!





Developmental Measures:

Since reporting of long-term outcomes began in 2011, through 2014, 100% of children with PKU age 0-12 receiving services in Nebraska were assessed by the pediatric metabolic specialists as meeting developmental milestones.

The following chart compares Nebraska PKU educational attainment with state & nat'l averages from the most recent year (2012) Nat'l Center for Higher Education. (Nebr. PKU 2014 #'s)

Measure	State Average	US Avg
% ages 25-64 with High school diploma	86.3% (Nebr. PKU 96%)	86.03
% ages 25-64 with Associates degree or higher	44% (Nebr. PKU 64%)	39.49
% ages 25-64 with a Bachelors degree	28.1% (Nebr. PKU 58%)	30.71
% ages 25-64 Graduate or Professional Degree	10% (Nebr. PKU 16%)	11.07

NEBRASKA EARLY HEARING DETECTION AND INTERVENTION ANNUAL REPORT - 2014

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

Approximately one to three in every 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 impact the child's speech and language acquisition, academic achievement, and social and emotional development. If detected soon after birth, the 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 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 and 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 information about 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

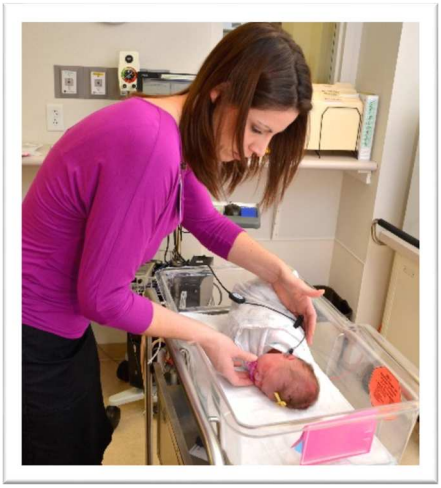
auditory, speech, and language developmental process in children. The Act also requires that regulations be promulgated to mandate newborn hearing screening if less than 95% of newborns in the state receive a hearing screening.

Newborn Hearing Screening Data Reported for 2014

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 2014, there were 56 birthing facilities conducting hearing screenings.

Hearing Screening at Birthing Facilities and Birthing Centers

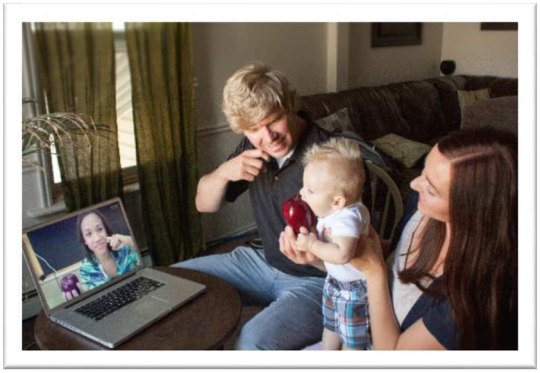


In 2014, inpatient hearing screens were reported on 26,858 newborns, or 99.7% of the 26,942 newborns available for an inpatient hearing screening. The percentage of newborns screened during birth admission has increased dramatically since reporting began in 2000, when only slightly more than one-third of newborns received a hearing screen during birth admission.

In 2014, 26,096 (97.2%) newborns passed the inpatient hearing 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 test 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 education materials free of charge to hospitals to help fulfill this requirement. Print materials are available in 10 languages.



Birthing facilities reported educating over 99% of parents about newborn hearing screening, hearing loss, and normal speech and language development in 2014. 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.

Monitoring, Intervention, and Follow-up Care

The NE-EHDI Program's tracking and follow-up procedures were followed for each baby who was reported as not passing the hearing screening during birth admission and for infants not receiving the inpatient hearing screening. In 2014, a total of 904 infants (hospital and non-hospital births) were tracked by the Nebraska EHDI Program to encourage the parent(s) to have the infant receive an outpatient hearing screening or audiologic evaluation.

The following shows the hearing screening/testing status of the 27,118 births for 2014:

26,096 Passed inpatient screening
707 Passed outpatient screening and/or diagnostic testing
113 Expired (inpatient or outpatient)
46 Parents refused screening and/or diagnostic testing
65 Diagnosed deaf or hard of hearing
64 Pending final screening or diagnostic testing
19 Lost (no response to NE-EHDI letters and phone calls)
8 Moved out of Nebraska

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, established by the Joint Committee on Infant Hearing (JCIH) of "1-3-6" (hearing screening completed by **1** month, audiologic diagnostic evaluation completed by **3** months, early intervention initiated by **6** months), the timeliness of initiation and completion of follow-up activities is an important aspect of the quality of services.

Over 97% of infants received an inpatient screening within one month of age. For the newborns who were recommended for an audiologic diagnosis, 67% received the evaluation by three months of age according to individual data received by the NE-EHDI Program from audiologists.

Records for the Early Development Network (EDN), Nebraska's Part C Early Intervention Program, indicated that 85% of infants residing in Nebraska in 2014, diagnosed as deaf or hard of hearing with a developmental delay, were enrolled in EDN services within six months of birth. The reasons for the 15% of infants not enrolling include: parents declining services,

unable to contact the family, family moved out of state, and no indication of developmental delay (diagnosed with slight or mild hearing loss).

ACTIVITIES – 2014

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 funded the basic operations of the NE-EHDI Program. The CDC Cooperative Agreement funded the development, implementation, and maintenance of the integrated electronic data reporting and tracking system.

In late 2013, a new three-year grant application was submitted for HRSA funding. Funding was approved and began April 1, 2014.

Advisory Committee

The NE-EHDI Program was created based on the requirements identified in the Nebraska Infant Hearing Act of 2000 and the recommendations of the NE-EHDI Program Advisory Committee. The purpose of the Advisory Committee is to provide direction and guidance to the NE-EHDI Program regarding the newborn hearing screening system. Specific advisory committee activities included, but were not limited to, the following:

- Discussed and advised on the goals for the NE-EHDI Program.
- Advised on the improvement of reporting, tracking, and follow-up protocols to effectively link the NE-EHDI Program and early intervention systems.
- Assisted in increasing the Program's responsiveness to the expanding diversity of cultural and linguistic communities in the state.
- Guided the long-term planning and evaluation of the NE-EHDI system in the state.
- Reviewed the quarterly newborn screening statistics and made recommendations for program improvements.

The advisory committee of the NE-EHDI Program included members representing the following:

- Audiologists
- Deaf/Hard of Hearing Community
- Early Intervention Services
- Ear, Nose, and Throat Specialist/Otorhinolaryngologists or Otologist
- Family Support Agencies/Organizations

- Hospitals (preferably hearing screening coordinator)
- Parents
- Pediatrics
- Public Health

Advisory committee meetings were held four times during 2014 and were open to the public.

Projects - 2014

HearU Nebraska/Children's Hearing Aid Loaner Bank

Formerly known as the Nebraska Children's Hearing Aid Loaner Bank, 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 2014. In 2014 there were 64 hearing aids provided to 37 children (age range of two months – 18 years) by HearU Nebraska. Since 2008 over 194 children have been provided hearing aids.

Roots and Wings Parent Weekend

The *Roots and Wings* parent weekend was held September 26-28, 2014 in Nebraska City at the Lied Lodge. The parent weekend targeted families with children up to 3 years old. The goal of this workshop was to provide: 1) families basic information on hearing loss, 2) an overview of current hearing technology, 3) knowledge on the various ways to communicate with deaf or hard of hearing individuals, 4) emotional support during the period after a family receives the diagnosis, and 5) an opportunity to network with other families. The positive participant survey results on the sessions and activities were shared in a presentation to the NE-EHDI Program Advisory Committee.

Parent Workshops

The NE-EHDI Program worked with Hands and Voices and the Regional Programs for Students Who Are Deaf or Hard of Hearing to organize workshops for parents of children who are deaf or hard of hearing. In 2014, workshops were held in Ashland and Kearney. The purpose of the workshops was to help parents build skills for effective parental advocacy for their child.

Website

The Nebraska Early Hearing Detection and Intervention Program website was created and can be found at http://dhhs.ne.gov/publichealth/EHDI/Pages/EHDIHome.aspx.

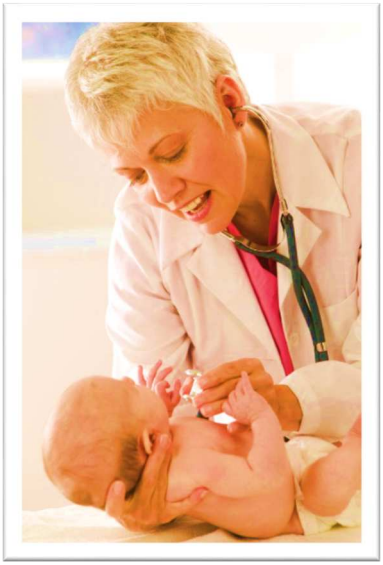
Task Forces

Two task forces were created during 2014. One began focusing on how hearing screening results are presented to parents, by birthing facility staff, when the baby does not does not pass the inpatient newborn hearing screening. The other is discussing issues related to cytomegalovirus (CMV) and the need for prevention, education, and possibly ways to integrate screening for CMV into hospital procedures screening.

Hospital Site Visits

During 2014, the program manager traveled Nebraska to visit 16 birthing facilities. The purpose of these visits was to determine what assistance the NE-EHDI Program could provide, how to lower refer rates, and to establish relationships with the hospitals.

Summary



- All of the 56 birthing hospitals in Nebraska were conducting newborn hearing screening in 2014, all but one conducted the hearing screenings prior to discharge from the hospital or birthing center.
- In 2014, birthing hospitals reported screening the hearing of over 99% of newborns prior to discharge from the hospital.
- The overall "refer" rate (those who do not pass) during 2014 for hearing screening during birth admission was 2.8% with over 85% of all newborns/infants completing recommended follow-up of outpatient screening/diagnostic testing. Seven percent were still in the process of completing the outpatient screening/diagnostic protocol in 2015.
- In 2014, confirmatory audiologic evaluations were completed within three months of age for almost 70% of newborns who did not pass the inpatient/outpatient screening or did not receive an inpatient screening.
- There were 137 infants born in 2014 whose hearing status remains unknown 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 4) families moving out of Nebraska.
- The incidence of Permanent Congenital Hearing Loss of 2.4 per thousand screened is within the anticipated range of one to three per thousand.
- Eighty-five percent of the infants identified as deaf/hard of hearing and residing in Nebraska were verified for the Early Development Network and received special education services within six months of birth.

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, drawn early specimens
Karen Eveans, NBS Follow up Specialist 402-471-6558
Hemoglobinopathies and cystic fibrosis, unsatisfactory specimens
Jessy Davis, Administrative Assistant 402 471-9731
Metabolic foods, patient education materials, advisory committee and staff support

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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:

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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, 402-471-3579
Follow-up, patient education materials distribution, data management
Courtney Smejdir, Community Health Educator, 402-471-6746
Follow-up, complex diagnostics, special projects
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Follow-up, community outreach, and education

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

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