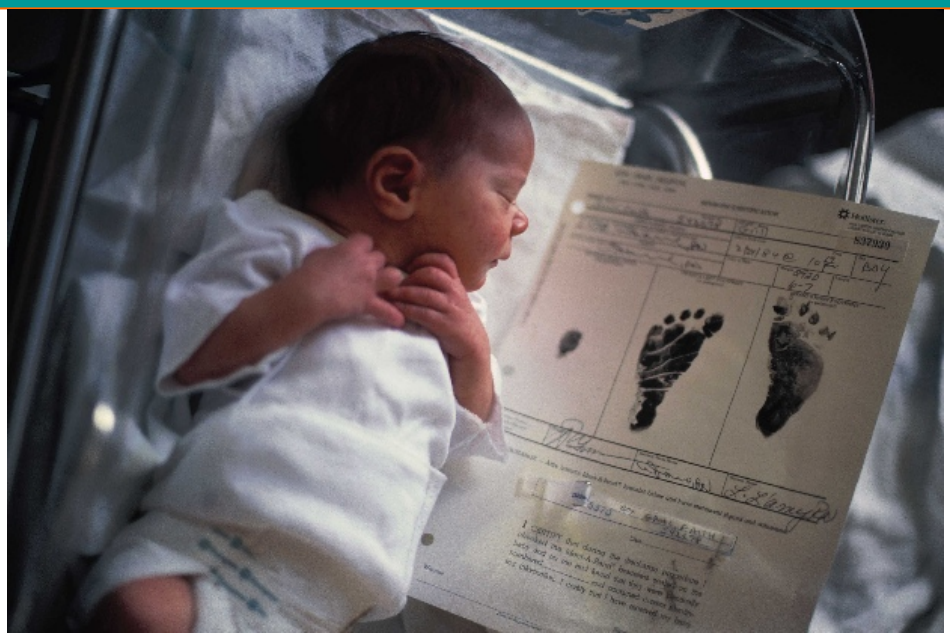
NEWBORN SCREENING IN NEBRASKA

Newborn Screening for Metabolic and Inherited Disorders

AND

Early Hearing Detection & Intervention



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NEWBORN SCREENING FOR INBORN ERRORS OF METABOLISM AND INHERITED DISORDERS

The goal of newborn screening for metabolic and inherited disorders is to identify newborns at risk for certain metabolic, endocrine, hematologic and other conditions that would otherwise be undetected until damage has occurred, and for which intervention and/or treatment can improve the outcome for the newborn.

Newborn Screening is a system 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 transmittal of specimens to the Newborn Screening laboratory
- Rapid quality testing methods
- Timely notification of the infant's parents
- Timely retrieval 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

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

In 2007, newborn screening efforts resulted in successfully identifying and treating 41 newborns affected with conditions in time to prevent problems associated with them:

- 4 babies with partial (treated) biotinidase deficiency
- 1 baby with congenital adrenal hyperplasia
- 16 babies with congenital primary hypothyroidism
- 12 babies with cystic fibrosis (11 classical, 2 atypical)
- 1 baby with duarte Galactosemia variant
- 4 babies with hemoglobinopathies (1 sickle hemoglobin-C disease, and 1 hemoglobin-C disease, 1 Beta Thalassemia Major, and one Sickle Beta Thalassemia)
- 2 babies with transient tyrosinemia (1 treated)
- 1 with methylalonic acidemia
- 1 with glutaric acidemia type I

The incidence rate of conditions in Nebraska based on the screened conditions identified in 2006 & 2007 (8 required plus supplemental) was 1:658.

WHAT IS NEWBORN SCREENING?

Newborn screening is the process of checking all newborns to identify those who are at greater risk of having one of the screened-for conditions. Newborn screening programs have been around for over four decades in all 50 states and in several other countries. The compulsory screening panel varies from state to state but the overall goal is the same: prevent or minimize the serious effects of these conditions. In 2007, the conditions each state screened for continued to became more uniform as an increasing number of states adopted screening by MS/MS (Tandem Mass Spectrometry). Nebraska has required this MS/MS supplementary screening to be offered to every newborn's parents since 2003.

The effects of not treating these conditions can include damage to the child's heart, kidneys, liver, spleen, eyes, nerve cells or brain. This damage can result in problems with growth, severe mental retardation, stroke and even death.

In 2007 Nebraska required screening for 8 conditions:

- **Biotinidase Deficiency (Bio)**
- **Congenital Adrenal Hyperplasia (CAH)**
- **Congenital Primary Hypothyroidism (CPH)**
- **Cystic Fibrosis (CF)**
- **Galactosemia (GAL)**
- **Hemoglobinopathies, (Sickle Cell Disease , Sickle hgb. C disease and thalassemias)**
- **MCAD**
- **PKU**

The conditions for which screening is done are individually rare, so consultation with and/or referral to the appropriate pediatric specialist such as a geneticist, metabolic specialist, hematologist, endocrinologist or an Accredited CF Center is always recommended.

In addition to the 8 required tests, Nebraska required that every parent be offered the option of "supplemental testing" for amino, fatty and organic acid conditions by tandem mass spectrometry.

Individually each condition is quite rare. However, collectively as many as one in every 625 babies are diagnosed each year in Nebraska with conditions from the current blood spot screening panel.

Adding hearing screening to these numbers, about 1:250 babies in the US are identified with screened conditions for which intervention is making a big difference in the lives of these children and their families.

In 2007, the program began the process to add the conditions in the supplemental screening panel to the required screening panel.

This action was taken in response to the recommendation of the US Department of Health & Human Services Secretary's Advisory Committee on Heritable Diseases in Newborns and Children, that all states screen for 28 core conditions.

The Nebraska March of Dimes and Nebraska Newborn Advisory Committee recommended to the Department regulation revisions to adopt this screening panel.

By the end of 2007, regulation changes were adopted and plans to implement July 1 of 2008 were underway.

HOW THE NEWBORN SCREENING PROCESS WORKS

1: TESTING

Baby is born.
Dried blood spot specimen is collected @ 24-48 hours of life.

Specimen shipped overnight to newborn screening laboratory, Pediatrix (now Perkin Elmer).

Specimen data entered into data system.

Specimen tested for multiple conditions.

2: FOLLOW-UP

Inconclusive or positive screen results reported by phone/fax/letter from lab and follow-up staff to baby's physician.

Baby's physician or health care provider contacts baby's parents.

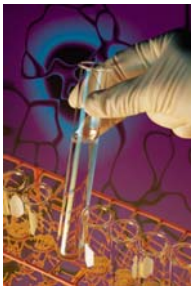
Parent's bring baby back in

for evaluation and more testing.

3: DIAGNOSIS/ INTERVENTION

Depending on the screen result, and on the condition screened:

Repeat or confirmatory testing occurs.



Parent education for signs/symptoms to watch for.

Baby's physician consults with and/or refers baby to pediatric sub-specialist appropriate to the condition.

4: TREATMENT & MANAGEMENT

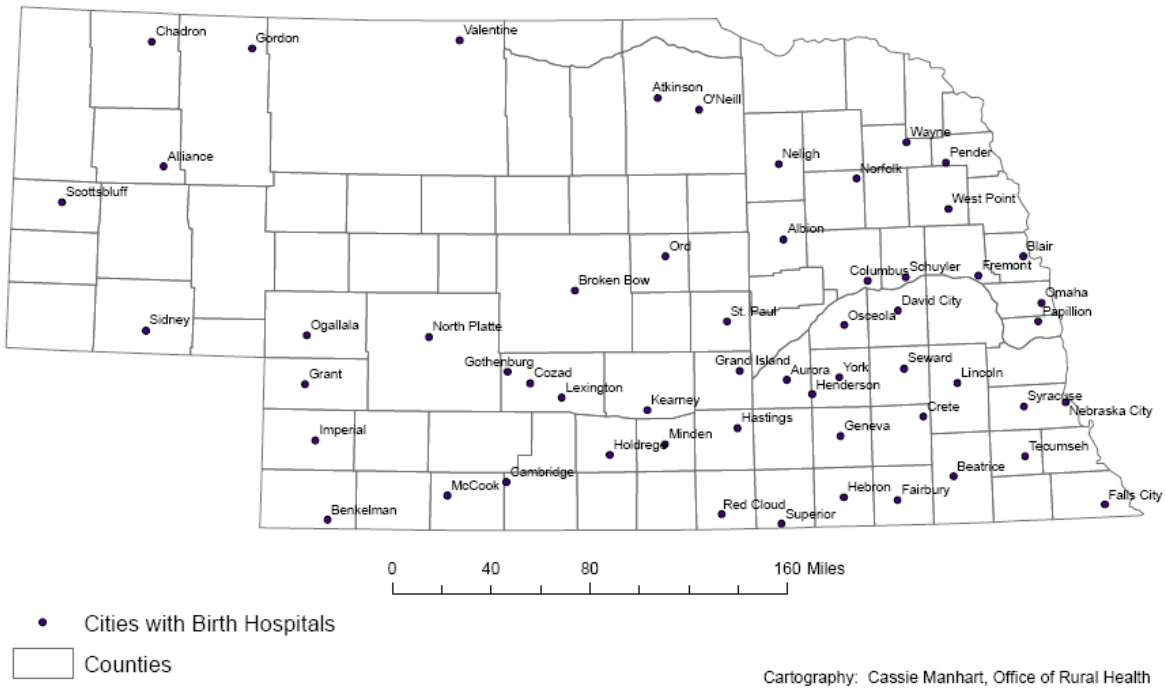
Once diagnosis is made, treatment begins. (For some life threatening conditions, treatment may occur prior to diagnosis on recommendation of specialist.

Parent's receive treatment guidelines / education.

Team Support services as appropriate, e.g.:

- metabolic dietitian monitoring & consultation
- ongoing blood monitoring
- referral to early intervention services
- pulmonary/ CF services
- ped endocrine monitoring
- ped hematology monitoring
- genetic counseling & consideration of family testing
- Other allied health services as needed

Nebraska Cities with Birth Hospitals



* 2 birth hospitals in Lincoln, 6 birth hospitals plus Children's Hospital in Omaha

System Overview

In 2007, 64 Nebraska hospitals sent specimens to Pediatrix Screening Laboratory. This laboratory is under contract with the State of Nebraska to conduct all of the newborn screens.

Supplemental screening (additional test results from Tandem Mass Spectrometry testing) was provided at no extra cost and required no extra blood. The supplemental screening provides results on fatty acid, amino acid and organic acid disorders. Educational efforts by physicians and hospital staff using written materials and videos provided by the Newborn Screening Program helped parents understand their options. More than 97% of parents consented to the supplemental screening for their newborns in 2007.

The Newborn Screening Program in the Nebraska Department of Health and Human Services was staffed by Mike Rooney, Administrative Assistant, Krystal Baumert, Follow-up Coordinator, Karen Eveans, Follow-up Specialist, and Julie Miller, Program Manager.

Ongoing consultation with the laboratory and specialists ensured expert advice and assistance was available as needed throughout the year. The program accessed metabolic specialists Richard Lutz, M.D., William Rizzo M.D., and Jill Skrabal, R.D., the Cystic Fibrosis Center Director John Colombo, M.D. and Dee Aquazzino and pediatric hematologist James Harper, M.D.

Quarterly meetings with the Newborn Screening Advisory Committee provided invaluable guidance to the program on several policy and quality assurance issues.

Treatment services received substantial support via the $10 per infant screened fee, State General Funds and Title V Maternal and Child Health Block Grant funds. This included funding for special metabolic formulas, metabolically altered/pharmaceutically manufactured foods, and support for specialty dietitian services and sub-specialist M.D. consultation services.

Quarterly quality assurance reports were sent to every birthing hospital, as well as Children's Hospital of Omaha, a facility that completes a significant number of screens on babies transferred to them. In addition, the Advisory Committee reviewed several quality assurance reports at each quarterly meeting.

The Egger Family Story:

My name is Jeanne Egger. I serve as a parent advocate on the Nebraska Newborn Screening Advisory Committee and the Heartland Genetics and Newborn Screening Collaborative. Two of my six children have Galactosemia.

Galactosemia is an inherited condition. It is a recessive genetic metabolic disorder. My two sons with the disorder do not have the necessary enzyme to digest the milk sugar, galactose. If untreated, it builds up in the body harming the eyes, the liver, the brain, and can be life threatening. Treatment is done by excluding foods with galactose from the diet. This is a diet for their entire life span. They do not outgrow it.

My husband, Dan and I were not aware of the potential for this disorder in our family. Our first two sons were what we expected. Lovely. But, they did have a bit more colic than what we thought was normal and just odd illnesses. Our family doctor advised us to see specialists when we discovered we were expecting another child, which we did. Nothing was found during the pregnancy. Robert was a healthy baby when we brought him home to join his brothers. He did have some jaundice and so we gave him sunbaths as advised to help him get over it. Finding a warm day in October can be iffy in Nebraska, but we managed to do that. However, his color worsened. He had been nursing normally at the hospital and at first at home. Then he started to throw up and then did not want to nurse at all. We ended up back at the hospital one week after he was born where Robert's pediatrician performed a complete blood transfusion. He went over the possible diagnoses with us. He mentioned several and said putting Robert on the milk-free diet would work with any of the possibilities that he had mentioned. Robert was transported from the neonatal intensive care unit in Lincoln to the University of Nebraska Medical Center in Omaha. The blood tests there showed that he had Galactosemia.

Our family had a new diet. Our relatives had a new diet and a reason to seek out genetic counselors. Our friends had a new diet when they came to our house. We read labels on every food item watching out for ingredients such as whey, casein, peas, and organ meats. Our neighbors called before they offered snacks to Robert and his little brother, Calvin. We kept milk free snacks at school for treat days and packed lunches. Eventually, Robert and Calvin started monitoring their own foods. They grew up, went to college and are currently serving in the military. Robert is married to his high school sweetheart and they have a beautiful daughter.

Screening for Galactosemia is now part of the panel recommended by the March of Dimes. Currently every baby born in Nebraska is tested by a few small blood samples taken from the heel. Our family was started before this screening was available. Our grandchildren get tested.

The Nebraska Newborn Screening Program would like to thank Jeanne Egger for sharing her family's story and for her many years of advocating for children through her service on the Advisory Committee and Regional Collaborative.

National Attention on Newborn Screening in 2007

Federal Legislation for Newborn Screening

For the first time, legislation at the Federal level specific to newborn screening and authorizing funds to support various aspects was introduced. Two versions of the legislation moved through the House and Senate during 2007. The bills amend the Children's Health Act and specify funding for the Secretary's Advisory Committee, grants to states, development of a national contingency/ emergency back up plan, the CDC's NBS QA/Proficiency testing program, and a national coordinating center and research. As of the printing of this report, the bill passed and was signed by the President in 2008.

A second piece of important legislation made progress moving through the House and Senate in 2007 related to genetics. Referred to as GINA, the Genetic Information Non-discrimination Act would provide further protections for individuals regarding how genetic information about them may be used.

Secretary's Advisory Committee Policy Recommendations

The federal Health and Human Services Secretary's "Advisory Committee on Heritable Disorders and Genetic Diseases in Newborns and Children" (SACHDGDNC) continued its efforts to evaluate the state of the states. The Committee developed a policy and procedure for evidence-based review and evaluation of candidate conditions for appropriateness for newborn screening as well as other recommendations to the Secretary of Health and Human Services. In 2007, proposals were submitted requesting approval for review of candidate conditions of Krabbe Disease, Fabry Disease, Spinal Muscular Atrophy (SMA), Severe Combined Immune Deficiency (SCID) and Fragile X.

The SACHDGDNC has four subcommittees established to assist in evaluating newborn screening systems and recommend priorities and strategies for insuring equity and quality among screening programs. The subcommittees are: Laboratory, Follow-up and Management, Education and Training, and Criteria Review Workgroup. Nebraska's own Amy Brower, Ph.D. of Third Wave Molecular Diagnostics served on the Committee and chaired the Lab Subcommittee. The Nebraska Program Manager had the honor of serving on the Follow-up Subcommittee. That subcommittee developed a paper on the elements of long-term follow-up and management, approved by SACHDGDNC and which was published in 2008. The Follow-up and Management Sub-committee also began evaluation of access to treatment, issues particularly focused on metabolic foods and formulas.

March of Dimes Advocacy

In 2007 the March of Dimes, Nebraska Chapter, continued to work with the Department to meet their national goal of ensuring every baby in every state received mandatory screening for all 29 conditions in the core panel. They testified at the public hearing in support of regulation revisions to change the supplemental testing to mandatory.

Clinical Laboratory Standards Institute (CLSI) Guidelines

The CLSI Immunoassay/Ligand Committee approved development of guidelines under a project proposed by Nebraska's Program Manager Julie Miller, in conjunction with Judi Tuerck, RN, MSN, formerly with the Oregon Newborn Screening System. The guidelines will address newborn screening for premature, sick and low birthweight NICU babies.

MAJOR INITIATIVES of 2007 in NEBRASKA

Education

- Mike Rooney of the Nebraska Newborn Screening Program continued to track and distribute the "Parents Guide To Your Baby's Newborn Screening" to the 64 birthing hospitals, Children's Hospital and upon request to some obstetric, family physician and pediatric practices.

- Following a survey to determine interest, the program distributed a parent education video "Newborn Screening, Protecting Your Baby's Health" to several local public health departments, federally qualified health centers, public health clinics, and obstetricians and family practice physicians providing obstetric services. The purchase was supported by the Heartland Regional Newborn Screening and Genetics Resource Center. The goal is to begin educating expectant parents about newborn screening prior to the birth of the baby.

- In February, Pediatric Grand Rounds at Children's Hospital and broadcast via satellite focused on updates in newborn screening. Featured speakers were Dr. James Harper, Pediatric Hematologist, Dr. G. Bradley Schaefer, Geneticist, Julie Miller, Newborn Screening Program Manager, Dr. Richard Lutz, Pediatric Geneticist/Endocrinologist, and Dr. John Colombo, Pulmonologist from the University of Nebraska Medical Center.

- In April, Dr. Richard Lutz presented a newborn screening talk at Saint Elizabeth Regional Medical Center in Lincoln.

- In June, Dr. James Harper presented newborn screening information at the Obstetrics Diagnostic Conference.

- Other invited talks were provided by the Program Manager to a parent group from the March of Dimes, and to Department of Health and Human Services representatives from the Northeast Service Area of Nebraska.

- Program staff participated in state-to-state collaboration via a newborn screening lab workshop held in Des Moines, and attended the Heartland Newborn Screening and Genetics Collaborative meeting in Oklahoma City. These meetings addressed quality assurance for newborn screening systems and issues related to the elements of lab testing, parent and professional education, follow-up and management.

- The Newborn Screening and EHDI programs displayed educational materials at an exhibit of the Missing Links Minority Health Conference held in Lincoln, which focused on reducing health disparities via appropriate language translation.

- The Nebraska Newborn Screening web-page was revised to address literacy levels.

- Internal staff development efforts included the program manager and follow-up staff attending the Association of Public Health Laboratory's National NBS and Genetics

Symposium in Minneapolis. Julie Miller presented as part of two panels, one on the use of the Performance Evaluation and Assessment Scheme (PEAS) and one on using national data for program assessment and improvement.

- On the recommendation of the National Newborn Screening and Genetics Resource Center, Dr. Huya Sirin, of the Ministry of Health in Turkey, visited Nebraska's Newborn Screening program for 4 weeks in November and December of 2007. All aspects of the organization and management of Nebraska's Newborn Screening System were shared, and she was exposed to many aspects of the Early Hearing Detection and Intevention system. Dr. Sirin had been given responsibility for the newborn screening system in Turkey, a country with approximately 1.5 million births per year. She is evaluating its system and capacity for expanding the number of conditions screened.

Dr. Hulya Sirin, Minstry of Health in Turkey (center), pictured with Julie Miller (left), and Chief Medical Officer and Director of the Division of Public Health, Dr. Joann Schaefer

Policy

Regulations revisions require tandem mass spectrometry panel
In 2007, the regulations were changed following public hearing to require screening for conditions formerly screened on the supplemental or optional panel. Those included conditions tested by tandem mass spectrometry in the acylcarnitine and amino acid profiles beyond just MCAD and PKU.

Competitive Bid for Laboratory Services Contract
In late 2007 the Request for Proposals was published inviting bidders to perform the newborn screening laboratory testing. The successful award may be renewed annually for up to 4 years. This competitive bid process therefore is required every 5 years.

Revisions to Lab Screening Protocols
In an effort to reduce the risk of a missed newborn with congenital hypothyroidism, the cut-off for TSH for specimens collected from babies greater than 7 days of age was revised. Thyroid stimulating hormone normally gets lower as babies get older. An update on this protocol change was sent to all birthing hospitals. It is expected that we will rarely need to implement this cut-off, as few babies with high TSHs will have their repeat specimen collected at > 7 days and have TSHs less than 20 but higher than 15.

Consent Regulations Reviewed
A group of stakeholders conducted the annual review of the regulations governing model consent for predictive genetic testing (one for supplemental newborn screening, one for prenatal predictive genetic testing, and one for other predictive genetic testing). No changes were recommended.

Legislation Introduced in 2007
The program monitored LB 250, which was a bill introduced by Senator Synowicki that would have allowed for a religious exemption to the required newborn screening, allowing parents to dissent from having their newborns screened. The Legislature's Health and Human Services Committee heard testimony on this bill, but no further action was taken on the bill in 2007.

Financing Newborn Screening
The Newborn Screening Advisory Committee had developed a Position Paper on Fiscal Sustainability for Nebraska's Newborn Screening System. As legislation to implement the Committee's recommendation for increased newborn screening fees has not been introduced, the program has begun evaluating other alternatives. An evaluation of amounts billed to Medicaid for newborn laboratory testing was undertaken. The program also worked with the metabolic clinic staff to promote evaluation of the amounts insurance companies were reimbursing for the metabolic formula. 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. Title V Block Grant funds support administrative aspects of the program (education, follow-up, program management and quality assurance). The State General Fund appropriation has stayed the same since 1997, and the Title V Block Grant appropriation to the state is below 1997 levels. The program continues to look for creative ways to make shrinking funds go further as costs increase.

Quality Assurance

Quarterly quality assurance reports were sent to each birthing hospital and Children's Hospital in Omaha. These reports include the individual hospital's quarterly measures and a state-wide comparison on each measure. In addition, the publication "QI Hints" is sent out with each quality assurance report to the person designated by the birthing hospital administrator.

Topics in 2007 included:

- Procedures for preventing discharge without screening
- How to avoid delaying newborn screening results
- Improving turn-around time for earliest test results
- New blood collection standards
- A reminder about transfer forms

Hospitals are encouraged to make the QI Hints available to all staff involved with parent education, specimen collection and handling, result reporting and tracking of screening results.

The program also began an in-depth evaluation of turn-around times, surveyed hospitals with the best turn-around times to help identify best practices. Hospitals in the lowest 25th percentile were sent letters informing them of their relative performance and providing them with a list of the "best practices" collected.

NEWBORN SCREENING ADVISORY COMMITTEE

The Newborn Screening Advisory Committee (NBSAC) provided technical expertise and policy guidance to the Nebraska Newborn Screening Program. Members commit at least a half day every three months to advise the state program. Several members provided extensive review and consultation beyond the committee meetings to help the program meet the recommendations of the larger Committee. The following summarizes this guidance:

Quality Assurance Reviews:

In 2007, the Committee continued to review quarterly quality assurance reports from the program. The Committee also monitored aggregate data received by the program on supplemental screening using Tandem Mass Spectrometry. Refer to Section III of this report for summaries of this data.

Quality and Technical Reviews:

The parent experts and medical experts of the Newborn Screening Advisory Committee were invaluable to the program in reviewing and making recommendations on newborn screening education, testing, follow-up, evaluation and financing.

Dried Blood Spot Testing for Genetic Causes of Hearing Loss:

The Advisory Committee and Early Hearing Detection and Intervention Advisory Committee made recommendations to the Department on the use of the dried blood spots to test for cytomegalovirus and other genetic causes of hearing loss. As a result, procedures were implemented to inform newborns' physicians of the availability of the blood spot testing once the newborns were confirmed with hearing loss. Blood spots requested before they are destroyed may be retrieved for this diagnostic testing.

Hemoglobinopathy Screening and Diagnosis Technical Reviews:

The Hemoglobinopathy Committee (subcommittee of the NBSAC) met to address Hemoglobinopathy screening and diagnostic issues. Members James Harper M.D. Pediatric Hematologist, Jeanine Kean, R.N. and Douglas Stickle PhD, Chemist met in October with staff of the NBS program (Krystal Baumert, Karen Eveans, Julie Miller and the Medically Handicapped Children's Program, Dr. Jeanne Garvin). The recommendations which were implemented by the program were:

- Revise parent information materials for patients with sickle cell trait in response to new research showing a small but statistically significant increased risk of venous thromboembolism among those with sickle cell trait.

All Hazards Preparedness Plan

In the fall of 2007, the Newborn Screening Program participated in TERREX-07, a state and local level functional simulation exercise of the emergency preparedness plans. The simulation was to evaluate the communication capacity of newborn screening results in the event state personnel were ordered to evacuate the State Office Building. Because of off-site infrastructure the program was able to meet the objectives given the scenario of Pandemic flu. Weakensses identified in the plan were corrected before the end of 2007.

The members of the NBSAC in 2007 were:

- **Khalid Awad**, M.D., *Neonatologist*, Neonatal Care PC, Omaha
- **Lawrence Bausch**, M.D., *Neonatologist,* Saint Elizabeth Regional Medical Center, Lincoln
- **John Colombo,** M.D., *Pediatric Pulmonologist, Director,* Nebraska Cystic Fibrosis Center, UMC, Omaha
- **Kevin Corley**, M.D., *Pediatric Endocrinologist*, Children's Hospital, Munroe/Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Jeanne Egger,** *Parent***,** Hallam
- **David Gnarra,** M.D., *Pediatric Hematologist,* Children's Hospital, Omaha
- **VICE CHAIR: James L. Harper**, M.D., *Pediatric Hematologist*, UNMC, Omaha
- **Kathryn Heldt,** R.D., *Dietitian*, Children's Hospital Metabolic Clinic, Omaha
- **Mary Kisicki,** R.N., *Parent*, Papillion
- **CHAIR: Richard Lutz,** M.D., specialist in *Pediatric Genetics, Endocrinology, Metabolism,* Munroe/Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Bev Morton,** *Parent,* Lincoln
- **Samuel Pirruccello**, M.D., *Pathologist*, Regional Pathology Services, UNMC, Omaha
- **Christine Reyes,** M.D., *Pathologist*, Pathology Center, Omaha
- **William Rizzo,** M.D.**,** specialist in *Pediatric Genetics, Endocrinology, Metabolism,* Munroe Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Kathy Rossiter,** MSN, *Certified Pediatric Nurse Practitioner*, Children's Hospital Metabolic Clinic, Omaha
- **Jill Skrabal,** R.D., *Dietitian*, Munroe Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Corri Stearnes,** *Parent*, Omaha
- **Douglas Stickle**, Ph.D., *Technical Director, Clinical Chemistry*, UNMC, Omaha
- **William Swisher,** M.D., *Pediatrician,* Lincoln Pediatric Group, Lincoln
- **B.J.Wilson,** M.D., *Neonatologist/Perinatologist*, Saint Elizabeth Regional Medical Center, Lincoln, March of Dimes Representative

Assurance of Treatment and Management of Conditions

How Treatment and Management Is Paid For:

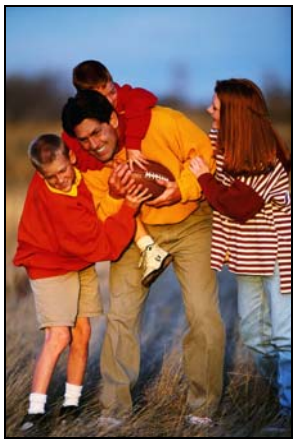
Part of the public health assurance role of newborn screening is ensuring treatment availability and access. Toward that end, the state program manages several contracts to ensure provision of otherwise prohibitively expensive formulas, foods, and services not always reimbursed by insurers. In 2007, 67 individuals were served through these programs: five infants, 35 children < 22 years of age, 4 pregnant women and 15 women of childbearing age, and 8 adult males > 21.

Insurance usually covers medical treatments for some screened conditions such as prophylactic penicillin for patients with sickle cell disease, or synthetic thyroid hormone for patients with congenital primary hypothyroidism. However, many do not cover the metabolic formulas, and none cover the pharmaceutically manufactured foods required for PKU and other metabolic conditions screened on the supplemental panel. Therefore, the biggest funding source supporting the metabolic foods and formulas was revenue generated from the $10 per infant screened fee (approximately $270,000 per year). The State General Fund appropriation of $42,000 also helped provide for these medically necessary formulas and foods and the associated nutritional counseling for patients identified with PKU or the other metabolic conditions identified on the supplemental screen. Title V Maternal and Child Health Block Grant funds then filled in the gaps for metabolic foods/formula and nutritional counseling. The Medically Handicapped

Children's Program provides some assistance to eligible families with children who have a hemoglobinopathy such as sickle cell disease or those with cystic fibrosis.

Individuals affected with screened metabolic conditions can obtain the metabolic formula through the Nebraska Medical Center Adult Metabolic Clinic, or at the Children's Hospital Metabolic Clinic. Ongoing dietary consultation, pediatric metabolic specialty care and routine blood monitoring are also provided. Individuals can order the pharmaceutically manufactured foods from product lists provided by manufacturers/distributors who have contracts with the state. Prior to 2007, 5 companies had contracts. In 2007, two more were added. Families can order up to $2,000 of the metabolically altered foods per year without having to pre-pay.

Nebraska's Families:


In Federal Fiscal Year 2007, metabolic formula ordering and distribution and specialized nutritional counseling and monitoring were provided via a contract with the University of Nebraska Medical Center for $302,817. Through this contract 67 patients with inherited metabolic conditions identified through screening were served at the metabolic clinics. During State Fiscal Year 2007, 50 of the 62 individuals eligible for the metabolic foods utilized the pharmaceutically manufactured foods program, ordering foods with a value totaling $56,465.90.



Mike Rooney coordinates the day-to-day metabolic foods program helping families to understand the program and stay connected, and monitoring the vendors' compliance with the contracts. He provides a tracking log to families for their use in monitoring their orders and expenses and provides a mid-year spending report to each family.

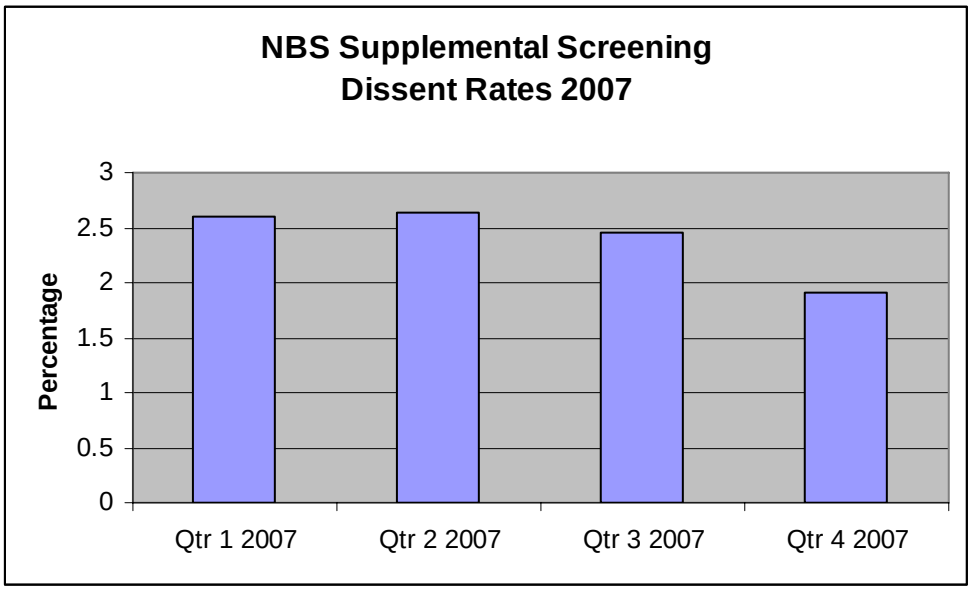
Sustaining The Obligation to Ensure Access to Treatment:

The number of children identified with conditions requiring special formula will always increase. The metabolic diets are required for life. People do not "age-out" of the need for the special formulas or foods. State General Funds have remained flat and federal allocations to Nebraska of Maternal and Child Health Title V Block grant funds have been reduced or flat for several years. While a new drug received FDA approval, for which about 40% of patients with PKU are expected to be responsive, these medications are expensive as well. Therefore the program continues to look for sustainable ways to continue to assure access to needed services for people who have these conditions.

PROCESS/OUTPUT DATA FOR 2007

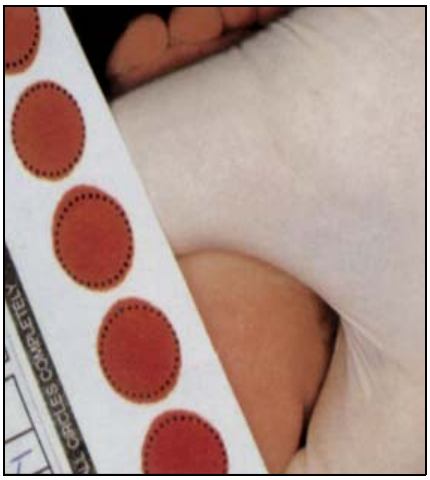
PATIENT EDUCATION

Consent for Supplemental Screening



Overall for 2007, 97.6% of parents consented to the supplemental newborn screening panel from MS/MS. By the last quarter of 2007, only 1.91% of parents dissented.

SPECIMEN COLLECTION, HANDLING AND TRANSPORT



Age at Time of Specimen Collection (Initial Specimen) 2007

Age at time of collection	Number of births	Percent of births
0-12 hours	198*	0.72
12-24 hours	133*	0.49
Collected day 2 (24-48 hours of age)	24,737	90.47
Day 3	1,958	7.16
Day 4	125	0.46
Day 5	34	0.12
Day 6	22	0.08
Day 7	18	0.07
Over 7 days	118	0.43

* 0-12 hours and 12-24 hours may include babies who had more than one specimen collected at < 24 hours, out-of-state births transferred to Nebraska NICU's, and possible duplication due to name changes.

Regulations require all specimens to be collected between 24-48 hours of birth, or prior to discharge, transfer or transfusion, whichever comes first. Specimens collected past day two are at increased risk of a delayed diagnosis.

Unsatisfactory Specimens for 2007

Number of specimens unsatisfactory / Total # initial specimens	114/ 27,013	0.42% of initial specimens
REASONS specimens were UNSATISFACTORY	**Number**	**% of unsats**
Blood spots not soaked through to the other side	33	29%
Quantity not sufficient	30	26%
Heavily applied, layered or double spotted	26	23%
Contaminated or diluted	8	7%
Exposed to heat or humidity	8	7%
Serum or fluid mixed with blood	7	6%
Sample got very wet prior to arrival in the lab	1	<1%
Unclear or conflicting patient identification on sample	1	<1%

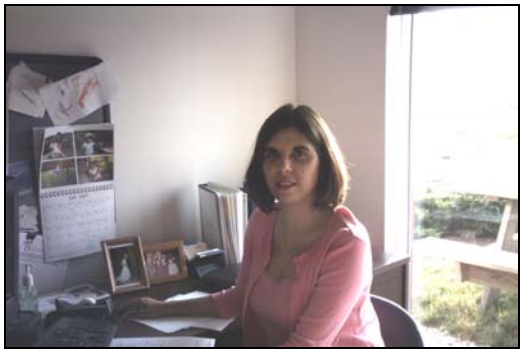
The art and science of correctly collecting and handling dried blood spots on filter paper requires trained health care professionals who consistently follow the Clinical and Laboratory Standards Institute procedures for specimen collection. Every unsatisfactory specimen must be repeated to ensure sufficiently reliable screening results.

Drawn Early (less than 24 hour) Specimens for 2007

Reason specimen collected at less than 24 hours of age	Number* / Percent
Baby to be transferred	180/265 or 68%
Baby to be transfused	12/265 or 4.5%
Unable to determine reason from data received at NNSP	73 or 27.5%

* Number is unduplicated, Nebraska-only births.

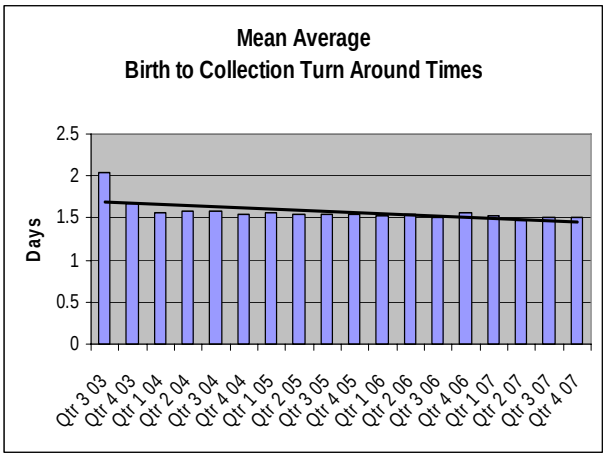
- Twenty-nine of the newborns whose specimens were drawn early did not get repeated as they expired. This was 20 more than the prior year.
- An additional 116 infants were reported as drawn early but when verifying with the birthing facilities they reported that they had made recording errors in these cases, and they submitted written corrections to the screening laboratory.

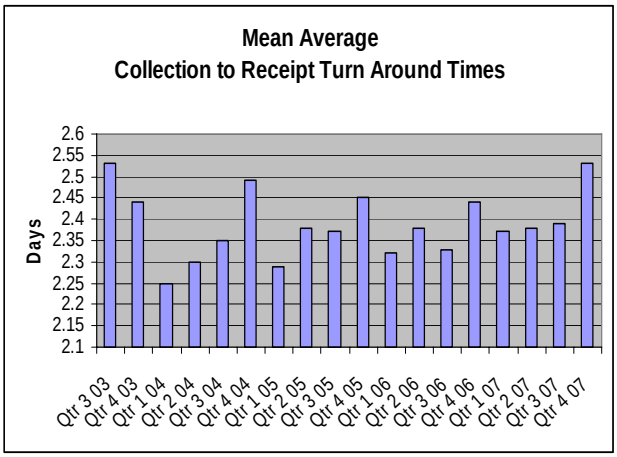


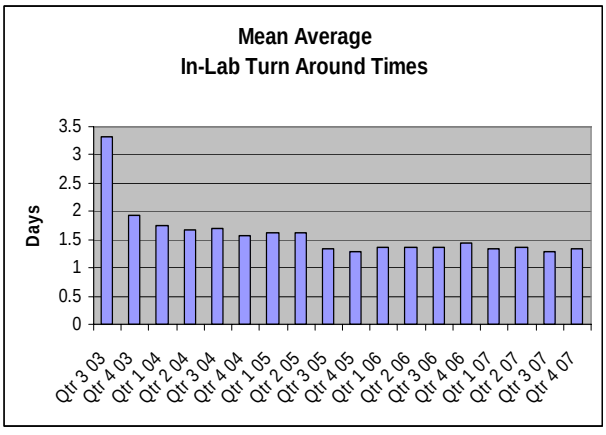
Helping to ensure rapid turn-around times are James DiPerna, Manager of MS/MS Laboratory Operations (above left), and Bethany Sgroi M.S.(above right), team leader for the 4 genetic counselors from Pediatrix screening laboratory, who quickly phone the positive or abnormal screening results to the Nebraska Newborn Screening Program, submitters and newborns' physicians.

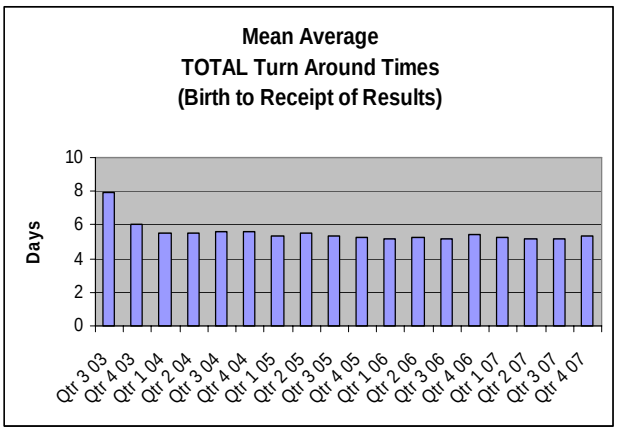
Specimen Turnaround Time

Regular monitoring of turn-around time between birth and reporting of results of the initial specimen is an important indicator for how well the newborn screening system is functioning. Specifically the ability to be able to turn specimens and results around in time to identify affected infants and prevent the effects of the condition. An initiative of the State Newborn Screening Program to more fully assess turn-around time resulted in changes of how data was presented to the Advisory Committee, and in some cases special reports to hospitals with turn-around times widely divergent from state averages. Likewise in-lab initiatives of Pediatrix screening laboratory resulted in reducing in-lab specimen processing/testing/reporting times as well. The following graphs identify statewide average specimen turn-around times over time. Of note are improvements in birth to collection, in-lab turn around and overall/total turn around times.

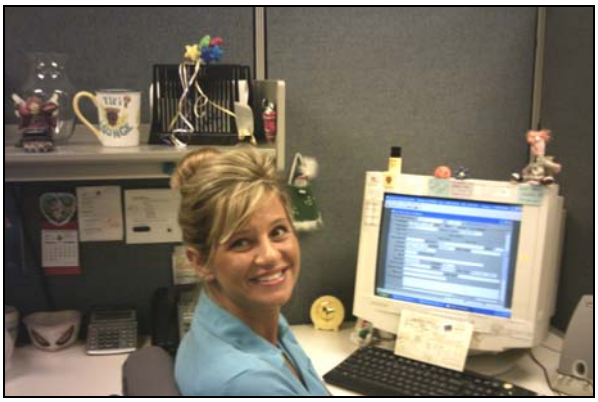








LABORATORY TESTING DATA



Heidi Oskamp, Pediatrix Screening laboratory, enters data into the Pediatrix electronic data system off of the tear-away sheets from the filter paper collection kits.

Pediatrix Screening laboratory staff perform testing on a variety of laboratory instruments.

Presumptive Positive Screening Rates

Screening programs by their very nature are designed to find those at higher risk of a disease in order to facilitate their diagnosis and treatment to prevent morbidity and mortality. Screening tests were never designed to be diagnostic, so a small percent of screen results will be positive that upon repeat or confirmation are found to be normal. Nebraska and programs across the country strive to minimize the number of newborns that require repeat or confirmatory testing (presumptive positive), and maximize the probability of identifying those affected. Nebraska continued to sustain a relatively low false-positive rate for every condition screened.

Including only the conditions required to be screened in 2007 (8), times the number of newborns screened (27,013), the number of tests completed for Nebraska newborns in 2007 were 216,104. Of this 216,104, there were 512 presumptive positive results requiring repeat or confirmatory testing. This is an overall presumptive positive rate of only 0.23%.

Over 97% of Nebraska newborns also received the supplemental Tandem Mass Spectrometry testing for the additional fatty acid, organic acid and amino acid disorders. Four of these required confirmatory testing and went on to diagnosis and treatment. Repeat dried blood spot specimens (not confirmatory testing) were needed for 234 of the supplemental screens as these were either mild elevations or were results affected by parenteral nutrition.

One of the many tandem mass spectrometers at Pediatrix screening laboratory.

Specific presumptive positive rates by condition		
Condition	**National rate 2007***	**Nebraska 2007 rates (mean average)****
Biotinidase Deficiency	0.04% 4:10,000	0.05% 6:10,000
Congenital Adrenal Hyperplasia	0.45% 45:10,000	0.08% 8:10,000
Congenital Primary Hypothyroidism	0.81% 81:10,000	0.22% 22:10,000
Cystic Fibrosis	0.49% 49:10,000	0.31% 31:10,000
Galactosemia	0.10% 10:10,000	0.03% 3:10,000
MCAD	0.02% 2:10,000	0.003% 3:100,000
Phenylketonuria	0.05% 5:10,000	0.003% 3:100,000

*National Rate 2007 is based on the sum of all reported presumptive positives divided by the sum of all the infants reported screened for the disease specified. This rate is converted from % to X:10,000 (rounded) for common reporting purposes. States with incomplete data e.g. (# positives not reported or consistent with # confirmed positive/negative and lost to follow-up) were eliminated from the data set. National data source: "2007 National Newborn Screening Report, Initial Screening Results," Biotinidase, Congenital Adrenal Hyperplasia, Congenital Hypothyroidism, Cystic Fibrosis, Galactosemia, MCAD, PKU newborns screened total column and newborns presumed with condition column. Number of states reporting: Biotinidase Deficiency 31, CAH 37, CH 37, Cystic Fibrosis 23, Galactosemia 37, MCAD 26 nd PKU 36 states reporting as of report run date of 09/04/08. Caution should be used in comparison of numbers.

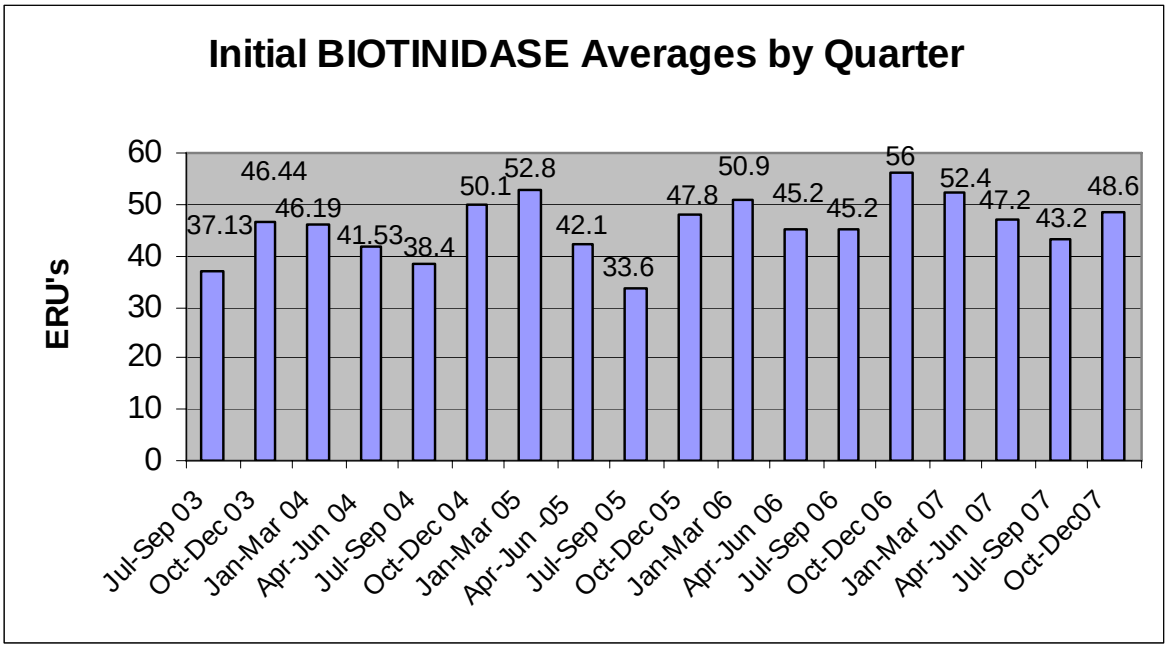
***Nebraska's rate 2007 is the number of presumptive positives divided by the total number of newborns screened in 2007. For MS/MS screened conditions, mild elevations of analytes requiring only repeat testing are not included. Significant abnormalities where confirmatory testing and referral to metabolic clinic are recommended are what was reported.

CAVEAT: States use varying instruments, methodologies and cut-offs. In addition, the national data report identifies inconsistencies in reporting by some states which brings into question the validity of the data. Therefore, direct correlations cannot be made from the data that are available. However, from the summary of data on the next page, one can extrapolate that, in general, Nebraska's chosen technology, methodologies and cut-offs have resulted in positive screening rates that are reasonable, compared to other newborn screening programs across the country. Rates for hemoglobinopathies were not figured due to variances in reporting methods for the national report, and from states. The national database uses data submitted by individual states, and can be found at http://www2.uthscsa.edu/nnsis/.

Mean Averages of Laboratory Test Measures

The program continues to provide lab testing data to the Newborn Screening Advisory Committee to monitor ongoing quality. The following tables depict the quarterly averages (mean values) for biotinidase measures, T_4 the primary screen for Congenital Primary Hypothyroidism, 17-OHP for congenital adrenal hyperplasia, Immunoreactive trypsinogen for CF and GALT and total galactose used to screen for Galactosemia. Access to data for mean averages for PKU and MCAD is not available from the Tandem Mass Spectrometry results from the screening laboratory. These means can tell us something about stability of the assay, reagents, etc., over time.

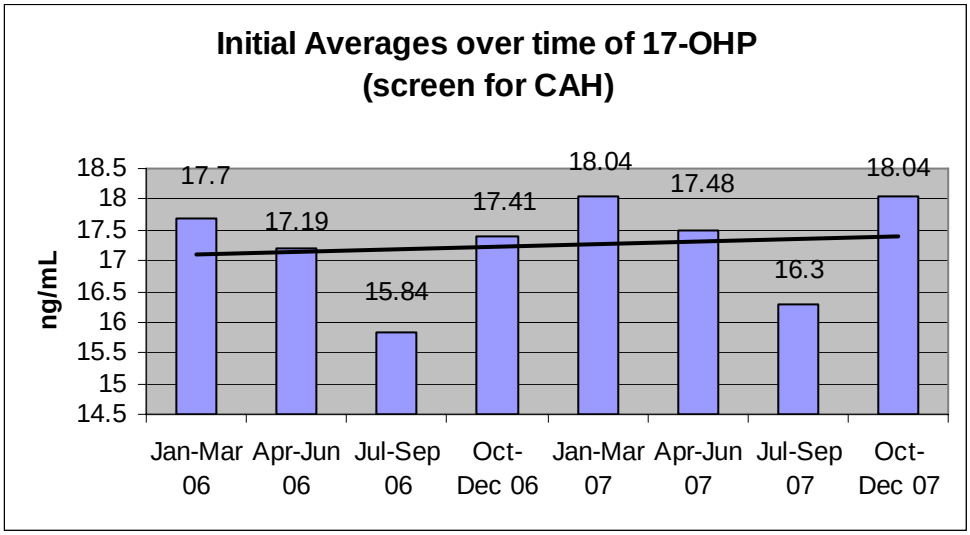
Health care providers familiar with these means might feel more comfortable explaining the "relative risk" to parents of newborns with positive screening results by comparing how far out of range the result is from the mean average and from the normal/expected range.

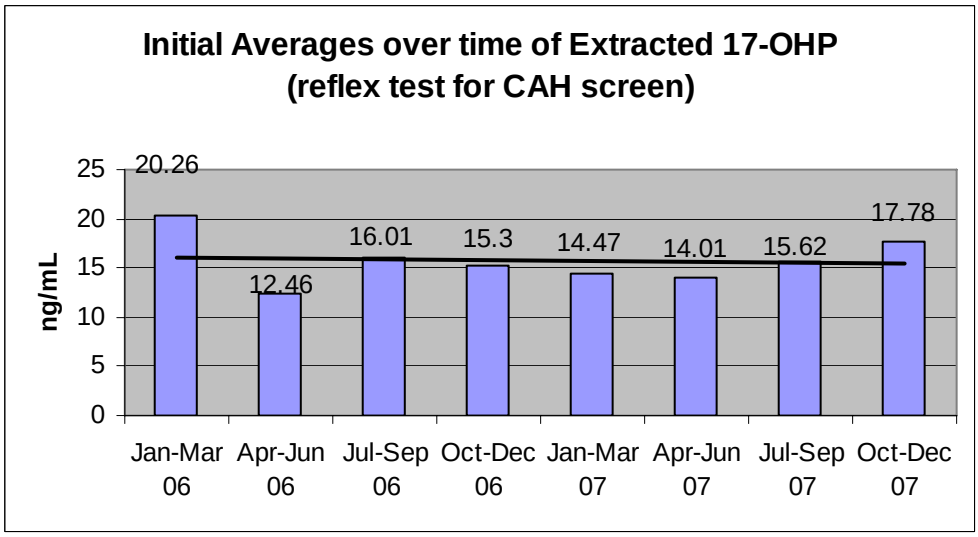


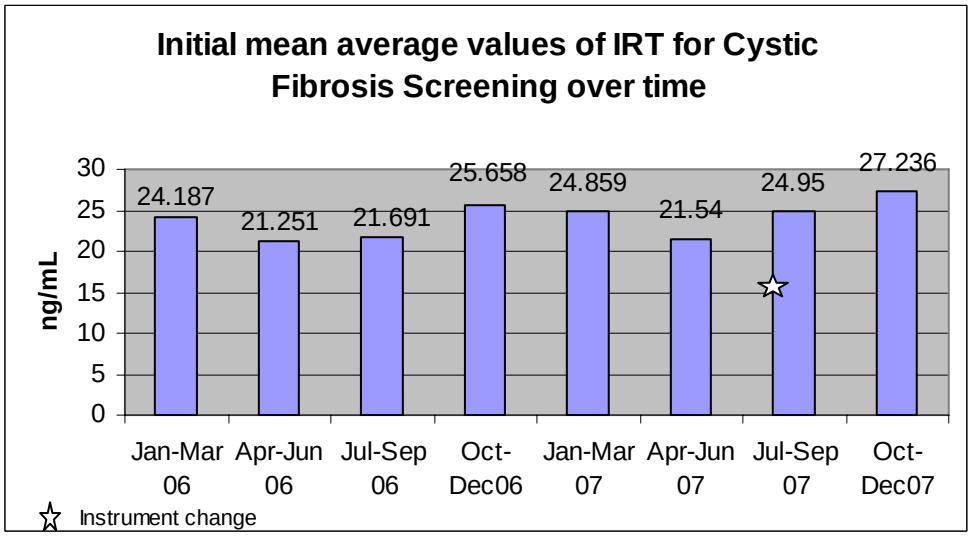
Expected seasonal differences can be seen each summer when heat exposure may impact the mean average enzyme levels detected in screening for biotinidase deficiency.

The Nebraska Newborn Screening Program sends a reminder each spring to hospital laboratory's about specific practices to follow that will minimize the risk of specimens becoming heat-denatured. This is intended to avoid the associated increase in the number of false-positive screens.

Reflex testing of abnormal CAH screens using an extracted 17-OHP reduces the number of positive screens reported and needing confirmatory testing.





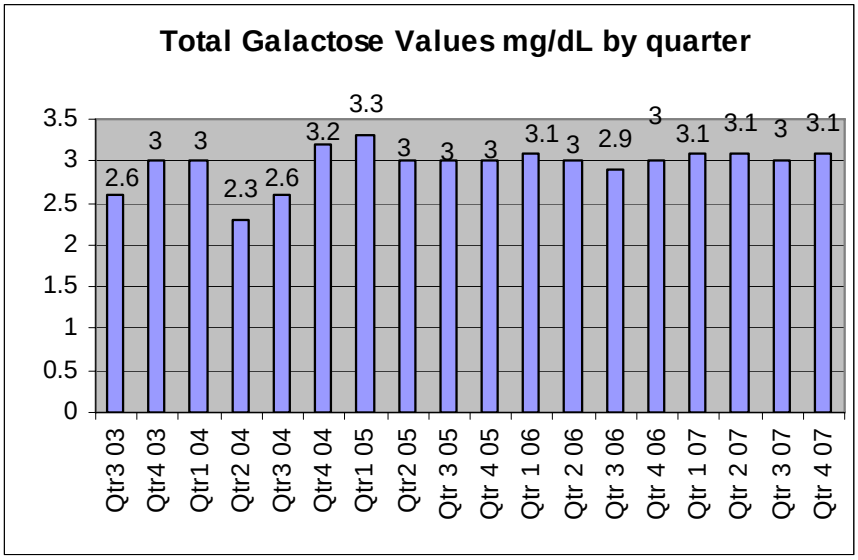


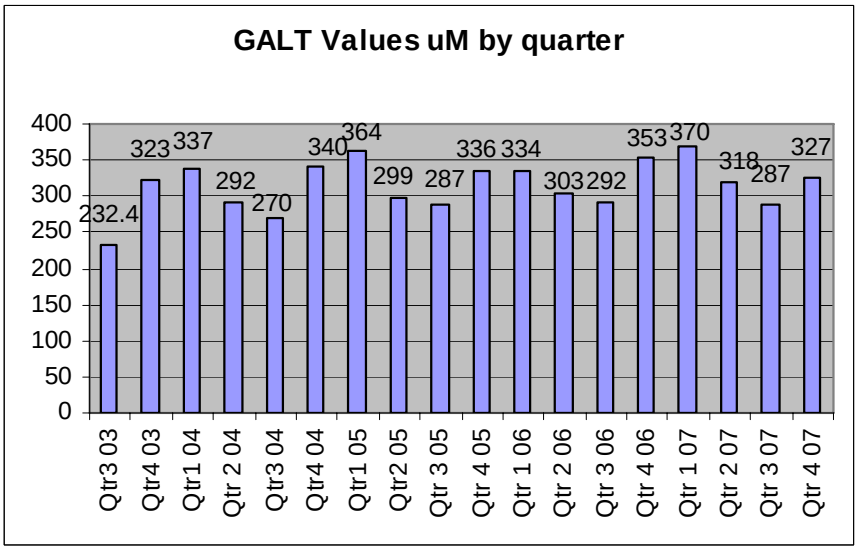
IRT's greater than 90 reflex to test for ΔF508 the mutation most commonly associated with classical cystic fibrosis. (Those with one copy of the ΔF508) automatically get tested for additional mutations on the INNOGENETICS 36 mutation panel).

By looking at both elevations of galactose and decreases in the enzyme activity of galactose phosphate uridyl transferase, the laboratory can report with greater precision those newborns at risk for classical Galactosemia who need immediate metabolic consultation/ referral and testing vs. those whose findings are more consistent with a milder but potentially clinically significant form of Galactosemia.

Having this information can mean providing more parents with a bit more peace of mind since most will need only a repeat screen vs. full confirmatory testing.

It can also mean cost savings by doing a "requested repeat" screen at no charge, vs. more expensive confirmatory testing.





Out of Hospital / Home Births

In 2007, there were 80 out-of-hospital births reported to the Department of Health and Human Services Newborn Screening Program (some reported later in 2008). All were screened, except for two of these babies who expired before they could be screened.

NEWBORN SCREENING DATA

	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007
Total Births	23,862	24,209	24,958	25,109	25,515	26,067	26,443	26,349	26,898	27,107 **
Births Screened	23,858 99.9%	24,118 99.9%	24,863 99.6%	25,043 99.7%	25,478 99.85%	26,008 99.77%	26,391	26,288	26,819	27,013
Total Births Lost to Follow-up	4	9	6 + (89 not screened -as expired @ <48 hours.)*	2 + (64 not screened as expired @ < 48 hours)	5 + (32 not screened as expired @ < 48 hours)	5 + (54 not screened as expired @ < 48 hours)	2 + (50 not screened as expired @ < 48 hours)	0 + (61 not screened as expired @ < 48 hours)	2 + (79 not screened as expired @< 48 hours)	
Total Births PP	547	357	412	432	456	415	499	503	537	511
Home Births	83	86	109	93	99	70	60	55	69	80
Home Births Screened	81	77	105	88	95	65	60	54	69	78
Home Births Lost to follow-up[1]	2	9	4	2 + (3 expired)	2 + (2 expired)	3 + (2 expired)	0	0 + (1 expired)	0	2 (both expired)

*Began match with death records beginning in calendar year 2000, to more accurately report #s actually screened.
** birth #'s are based on births reported to the NNSP and matched with vital records birth system ending 4/15/2007. This differs from the Early Hearing Detection and Intervention data because of a later cut-off used for EHDI data.

Biotinidase Deficiency	**1998**	**1999**	**2000**	**2001**	**2002**	**2003**	**2004**	**2005**	**2006**	**2007**
Presumptive Positive	3	4	2	4	3	4	34*	78	14	5
Inconclusive										10
Confirmed Negative	2	2	2	1	1	0	29	71	9	11
Confirmed Positive Profound	1	1	0	0	2	1	0	1	0	0
Confirmed Positive (Partial no tx)	0	0	0	0	0	0	0	0	0	0
Confirmed Positive (Partial tx)	0	1	0	3	0	3	6	5	4	4
Lost to follow-up	0	0	0	0	0	0	0	1**	1**	0

*Screening protocols identified most of these as "inconclusive," for which repeat screening rather than confirmatory testing ruled out the condition.
** lost to follow-up as newborn expired

Congenital Adrenal Hyperplasia	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007
Presumptive Positive	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	10	3
Inconclusive										18
Confirmed/ repeated Negative	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	9	17
Confirmed Positive	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	1	1
Confirmatory or Repeat Lost to follow-up	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	0	3*

* expired before repeat or confirmatory testing could be done. Includes one set of twins.

Congenital Primary Hypothyroidism	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007
Presumptive Positive	274	108	114	115	129	89	63	58	51	40
Inconclusive (drawn early but low T4/high TSH										22
Confirmed Negative	265	92	104	105	113	75	55	48	41	43
Confirmed Positive	6	13	8	7	15	11	8	9	10	16
Confirmatory or Repeat Lost to follow-up	3	3	2*	3*	1*	3*	0	1*	0	3*

*Lost to follow-up as babies expired.

Data for Cystic Fibrosis (below) and Hemoglobinopathies (next page) are presented in different format because screening for CF is inherently more complex, and diagnosis for hemoglobinopathies can be more protracted and complex. Although the goal is to detect clinically affected newborns to initiate early treatment and prevent infant mortality and morbidity, the screening test can detect some carriers or people who have the trait for these conditions.

Cystic Fibrosis:	**Year**	**2006**	**2007**
Total Screened Positive		8	4
Of those:	Confirmed CF	8	4
	Confirmed Atypical CF	0	0
Total Screened Inconclusive		62	54
Of those:	Confirmed CF	2	3
	Confirmed Atypical CF	2	0
	Confirmed Carriers	10	12
	Found to be within normal limits on repeat	46	35
	Expired before confirmation could be done	1	4
	Lost to follow-up	0	0
	Pending	1	0
Total with Meconium Ileus or Bowel Obstruction and positive on DNA		4	13
Of those:	Confirmed CF	1	5
	Found to be within normal limits	3	7
	Pending diagnosis	0	1

Galactosemia	**1998**	**1999**	**2000**	**2001**	**2002**	**2003**	**2004**	**2005**	**2006**	**2007**
Presumptive Positive	9	13	12	15	5	3	9	1	8	0
Inconclusive repeat rec'd										9
Confirmed / repeated Negative	9	8	8	9	5	0	6	1	8	8
Confirmed Positive (Classical)	0	0	1	0	0	1	0	0	0	0
Confirmed Positive, Duarte (not treated)	0	3	1 Duarte Hmzgt	0	0	1	0	0	0	0
Confirmed Positive, Duarte (treated)	0	2	2 Duarte Mixed Htrzgt. (1 tx'd 1 year)	6 Duarte Mixed Htrzgt.	0	1	3	0	0	1

Hemoglobinopathy Follow-up Changes:

In 2007, follow-up procedures added the step of sending a reminder letter before the 6-month checkup if the initial confirmatory report indicated a possible alpha, beta or gamma chain variant (or combination in the heterozygous state) alpha thalassemia or beta thalassemia. These typically require additional blood work to diagnose, which previously was not usually reported back to the program. This has resulted in a significant increase of diagnosed and closed cases. Ultimately, the goal is to provide families with better information about their child's hemoglobinopathy.

Abbreviation Key (likely diagnosis associated with screening results)

FS:	Sickle Cell Disease	FAS:	Sickle Cell Trait
FC:	Hemoglobin C Disease	FAC:	Hemoglobin C Trait
FSC:	Sickle Hemoglobin C Disease	FAD:	Hemoglobin D Trait
FE:	Hemoglobin E Disease	FAE:	Hemoglobin E Trait
FSA:	Sickle Beta Thalassemia	FAV:	Hemoglobin Trait - unknown variant
HPFH:	Hereditary Persistence Fetal Hemoglobin		

Clinically Significant Hemoglobinopathies Confirmed Positive:

	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007
FS	1	3	2	4	4	5		1	3	
FC							1	1		1*
FSC	1	1	1	2	2		1	1	2	1
FE			1						1	
Sickle Beta Thal										1
Alpha Thal Major				1 (4-gene deletion)						
Beta Thal Major										1
HPFH								1		
FAE + possible Beta thal									6	
FAS + possible Beta Thal									11	
FAS + possible Alpha Thal									9	
FAC + possible Alpha Thal									3	

Dx. = Hemoglobin C Disease or Hemoglobin C beta thalassemia

Other Hemoglobinopathies Confirmed Positive in 2007:

141	Sickle Cell Trait	30	alpha Thalassemia trait
37	Sickle hemoglobin C Trait	3	alpha Thalassemia silent carrier
8	Hemoglobin D Trait	15	Sickle cell trait plus other
11	Hemoglobin E Trait	6	Sickle cell trait plus alpha Thalassemia trait
4	Other Variant/Trait	3	Sickle Cell trait plus alpha Thalassemia silent carrier
6	Hemoglobin C trait plus other	2	Hemoglobin E trait plus other
1	Gamma Chain Variant		

Diagnosis unknown for 105 positive hemoglobinopathy screens (not suspected for clinically significant conditions) (confirmatory results not reported back to program)

MCAD *	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007
Screened Positive	N/A	N/A	N/A	N/A	3*	3	5	10	5	0
Screened inconclusive (repeat only)**	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	2
Confirmed Negative	N/A	N/A	N/A	N/A	2	3	1	7	5	2
Confirmed Positive	N/A	N/A	N/A	N/A	1	0	4	3	0	0

*Mandatory screening for MCAD began 7/01/2002. Prior to that about 34% of newborns were voluntarily screened in Nebraska in 2000 and 2001.
**Inconclusive screen: Abnormal screen result requiring only a repeat screen, not confirmatory testing.

Phenylketonuria (PKU)	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007
Presumptive Positive	43*	3	6**	4	3	7**	7	3	6	0
Screened Inconclusive (repeat only)***	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	2
Confirmed Negative	40	0	2	2	1	1	1	1	1	2
Confirmed Positive Classical PKU	2	1	1	1	1	2	1	2	0	0
Confirmed Positive Hyperphe		2 (tx'd)	1 Transient	1	1	3	5 (3 of these tx'd)	0	5 (4 of these treated)	0

1998: One confirmatory testing not done – residence in another state
****2000 and 2003***: One each year for whom confirmatory testing was not done as the babies expired
***Inconclusive screen: Abnormal screen result requiring only repeat screen, not confirmatory testing.

Tandem Mass Spectrometry Supplemental Screening Results

SUMMARY OF MS/MS FINDINGS Jan 1 – Dec 31, 2007

(Including MCAD and PKU, MS/MS screened conditions on the required screen)

Numbers include a few babies with one abnormality on screen and a different abnormality on repeat.

Initial findings	Out of Range on Screen	Repeated or Confirmed negative	Pending or Lost to Follow-up	Confirmed Positive
C3 (Propionylcarnitine)	15 (+ 1 on repeat, + 1 out of state birth)	16	(1 out of state birth lost to follow-up)	
C3 & C3/C2 & C3/C16 (Propionylcarnitine and ratios with acetylcarnitine and palmitoylcarnitine)	25 (+ 2 on repeats, 1 out of state birth & 1 out of country birth)	28		1 Methylmalonic Acidemia
Several Amino Acids (likely hyperalimentation)	75 (+ 1 out of state birth + 10 on repeat screen)	74	11 (expired) (+ 1 out of state birth expired)	
Several Amino Acids and Isovalerylcarnitine (C5) and other indices such as the relataive ration of C5 to butyrlcarnitine (C4)	1	1		
Methionine	74 (+ 18 on repeat screening, + 1 out of state birth)	85	7 (expired) (+1 out of state birth lost to follow-up)	
Tyrosine	20 (+5 on repeat)	23		2 Transient Tyrosinemia
Methionine and Propionylcarnitine (C3) and C2:C3 and C3:C16	1	1		
Glutarylcarnitine (C5DC)	3	2		1 Glutaric Acidemia Type I
Citrulline & Glutarylcarnitine (out of state birth)	(1 out of state birth)	(1 out of state birth)		
3-hydroxyisovalerylcarnitine (C5-OH)	2 (+ 1 on repeat)	3		
Methionine & Phenylalanine ratio	4	3	1 lost to follow-up out of country	
Methionine & Tyrosine	4	4		
Phenylalanine & Tyrosine Ratio	1	1		
3-hydroxybutyrylcarnitine (C4OH)	1	1		
Tetradecaanoylcarnitine C14:1	1	1		
Tetradecanoylcarnitine (C14:1) ratio to Palmitoylcarnitinne (C16) & other long chain carnitines	3	3		

Alanine	2	1	1 (expired)	
Butyrylcarnitine (C4)	4 (+1 on repeat)	5		
Butyrylcarnitine (C4) and C3 with C3/C2 & C3/C16	1	1		
Octanoylcarnitine	1 (+ 1 on repeat)	2		
TOTALS Babies*** Abnormal screens	 244 283	 256	 23	 4

*Lost to follow-up designated when the patient/parent can no longer be found and there is no medical home, or they have moved out of state to an unknown location.

**The vast majority of abnormal screens from MS/MS require only a repeat screen to rule out the condition. Confirmatory testing is recommended in a small percentage of cases where the concentration of analytes are "significantly" abnormal, or concentrations of analytes increase on repeat screens.

***Total babies less than # of abnormal screens as some that had more than one abnormal screen, e.g. methionine on initial screening, and multiple amino acids on a repeat screen.

Intervention Data

Intervention data is one of the most important measures for determining how well we are doing as a system to ensure timely treatment of affected infants.

The following data is grouped by condition and shows Nebraska's averages/ranges for 2007. In some cases "intervention" (family consultation, evaluation, and monitoring of the newborn) occurred well before the age actual treatment was initiated, as treatment was pending confirmatory testing and diagnosis.

The following data also includes national averages/ranges pulled from the "National Newborn Screening Report -2007" available at the National Newborn Screening and Genetics Resource Center's Web site. Data for this section of the Annual Report was run on 7/24/07.

http://genes-r-us.us.uthscsa.edu/resources/newborn/00chapters.html

Comparisons should be made with extreme caution. States and territories included in the averages in this report have reported fewer than 4,000 babies screened per year to around 500,000 per year. Likewise, resources necessary to complete testing, follow-up, confirmation, diagnosis and treatment also vary from state to state. The intervention data is one kind of outcome data that can, over time, help identify how well a state's system is working in newborn screening. The mean average age at time of treatment can be an indicator of whether adequate resources are devoted to each of the components of a comprehensive newborn screening system: education, specimen collection handling and transportation procedures, laboratory procedures, follow-up and referral procedures, confirmation and treatment.

Biotinidase Deficiency

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data
Goal age for treatment initiation: Upon Diagnosis	32 states reported data
# diagnosed/treated: 0 profound 4 partial deficiency's treated	46 cases of profound biotinidase deficiency reported
Mean avg. age at Initiation of treatment: 20 days	6 cases or 13% treated by 7 days of age 13 or 28% treated between 8-14 days of age 10 or 21% treated between 15-21 days of age 11 or 24% treated at > 21 days of age 6 or 13% age of treatment unknown/not reported
Age ranges at Initiation of treatment: 13-25 days	Age ranges at Tx initiation.: 5 - > 21 days

Source: http://www2.uthscsa.edu/nnsis/ data entered by 9/10/08

Congenital Adrenal Hyperplasia

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data
Goal age for treatment initiation: Upon Diagnosis	41 states reported data
# diagnosed/treated: 1	133
Mean average age at Initiation of treatment: 8 days	52 cases or 39% treated by 7 days of age 46 cases or 35% treated by 8-14 days of age 11 cases or 8% treated by 15-21 days of age 12 cases or 9% treated at > 21 days of age 12 or 9% age of treatment unknown/not reported
Age ranges at treatment initiation (N/A)	Age ranges at initiation of treatment: <3 - > 21

Source: http://www2.uthscsa.edu/nnsis/ data entered by 9/10/08

Congenital Primary Hypothyroidism

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data
Goal age for treatment initiation: As early as possible, upon diagnosis	47 states reported data
# diagnosed/treated: 14	# diagnosed/treated: 1645
Mean avg. age at Treatment initiation: 15 days	Age at initiation of treatment: 422 cases or 26% treated by 7 days of age 542 cases or 33% treated between 8-14 days of age 221 or 13% treated between 15-21 days of age 372 or 23% treated at > 21 days of age 88 or 5% age at tx. unknown or not reported
Age ranges at treatment initiation: 4-50 days	Age ranges at treatment initiation < 3 - > 21 days

Source: http://www2.uthscsa.edu/nnsis/ data entered by 9/10/08

Cystic Fibrosis

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data
Goal age for treatment initiation: As early as possible, upon diagnosis.	33 states reporting data
# diagnosed/treated: 12 Classical CF	# diagnosed/treated: 284
Mean average age at initiation of treatment: 18 days	Age at initiation of treatment: 57 cases or 20% treated by 15 days 49 or 17% treated at 16-30 days 30 or 11% treated at 31-45 days 16 or 6% treated at 46-60 days 14 or 5% treated at 61-75 days 6 or 2% treated at 76-90 days 7 or 3% treated at > 90 days 105 or 37% age of tx. unknown or not reported
Age ranges at initiation of treatment: 2-94 days (1 @ 94 days -delay due to social situation and dx. requiring observation & DNA sequencing). 5/12 presented with meconium ileus	Age ranges at initiation of treatment 1->90 days

Source: http://www2.uthscsa.edu/nnsis/ data entered by 9/10/08

Galactosemia

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data
Goal age for treatment initiation: As early as possible, upon diagnosis. Diet intervention upon positive screening result	38 states reporting data
# diagnosed/treated: 0 1 Duarte Variant Galactosemia treated at 6 days	70 cases of classical galactosemia identified
Mean avg. age at treatment initiation: N/A	Age at treatment: 34 or 49% treated at 7 days of age or less 14 or 20% treated between 8-14 days of age 3 or 4% treated between 15-21 days of age 8 or 11'5 treated at >21 days of age 11 or 16% age of treatment unknown or not reported
Age ranges at initiation of treatment: N/A	Age ranges at treatment initiation: < 3 days - > 21

Source: http://www2.uthscsa.edu/nnsis/ data entered by 9/10/08

MCAD - Medium Chain Acyl Co-A Dehydrogenase Deficiency

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data
Goal age for treatment / intervention initiation: As early as possible, upon positive screening result – parent education/ consultation.	41 states reported data
# diagnosed/treated: 0	# diagnosed/treated: 167
Average age at intervention (avoid fasting): N/A	Age from birth to treatment: 62 or 32% 7 days or less 50 or 26% between 8-14 days of age 18 or 9% between 15-21 days of age 27 or 14% at > 21 days of age 34 or 18% age treatment unknown
Age ranges at initiation of treatment: N/A	Age ranges at treatment initiation: <3 - > 21 days

Source: http://www2.uthscsa.edu/nnsis/ data entered by 9/23/08

PKU - Phenylketonuria (Classical PKU)

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data
Goal age for treatment initiation: As soon as possible but no later than 7-10 Days after birth.*	43 states reported data
# classical PKU: 0	159 cases of classical phenylketonuria
Avg. age at treatment: N/A	53 or 33% treated by 7 days of age 61 or 38% treated between 8-14 days of age 18 or 11% treated between 15-21 days of age 9 or 6% treated at > 21 days of age 18 or 11% age at treatment unknown or not reported
Age ranges at initiation of treatment: N/A	Age ranges at treatment initiation: < 3 - > 21 days

*NIH Consensus Statement October/25/2000: Phenylketonuria: Screening and Management
Source: http://www2.uthscsa.edu/nnsis/ data entered by 7/24/07

Hemoglobinopathies

Nebraska 2007 Intervention Data	U.S. 2007 Intervention Data[1]
Goal age for treatment initiation:[2] 60 days of age or less	40 states reported data
# cases diagnosed/treated Sickle Cell disease (S/S) 0 Sickle Hgb. C disease (S/C) 1 Beta Thalassemia major 1 Hgb C disease/ C Beta thal 1 Sickle Beta Thalassemia 1	# cases diagnosed/treated Sickle Cell Disease (S/S) 859 Sickle Hgb. C Disease (S/C) 455 Homozygous Hgb. (F only) 29 Hgb C disease/C Beta Thal 46 Sickle Beta Thalassemia 94
Mean/Average age (days) at treatment: For all clinically significant hgb's: 15 days	Due to space limitations the various breakdowns of ages at diagnosis for each type of Hemoglobinopathy are not listed here. Please reference the National NBS & Genetics Resource Center at genes-r-us@uthscsa.edu
Age ranges at initiation of treatment: 8-17 days 100% treated by 60 days of age.	Age ranges at treatment initiation: 0 - > 90

Source: http://www2.uthscsa.edu/nnsis/ data entered as of 7/24/07

[2] Treatment guideline from A Clinical Practice Guideline #6, Sickle Cell Disease: Screening, Diagnosis, Management and Counseling in Newborns and Infants, U.S. Dept. Of Health and Human Services, Public Health Service, Agency for Health Care Policy and Research.

Other metabolic conditions diagnosed through screening

Nebraska 2007 Metabolic condition / # detected / age @ treatment	U.S. 2007 (Save data but only for those conditions also found in 2007 in Nebraska, not all metabolic conditions found).
Glutaric Acidemia I 1 treated @ 3 days	22 cases (27% by 7 days, 23% by 14, 31% > 21)
Methylmalonic acidemia Cbl. A/B 1 treated @ 15 days	12 cases (8% by 7 days, 42% by 14, 25% by 21, 8% > 21 days)
Transient Tyrosinemia 1 treated @ 9 days	Not reported in national data system
Transient Tyrosinemia 1 not requiring treatment	Not reported in national data system

PLANS

Screening Panel Expansion: Nebraska screened nearly 100% of newborns* for eight conditions and greater than 97% of newborns for the additional organic acid, fatty acid and amino acid Disorders that can be detected on Tandem Mass Spectrometry screening. The regulation revisions requiring screening for those conditions on the supplemental screening panel that are screened by tandem mass spectrometry were implemented in 2008.

* Some babies die before the screening specimen can be collected.

Other System Improvement Planning Efforts: The Nebraska Newborn Screening Advisory Committee will continue to advise the Department of Health and Human Services on implementation of elements of the National Newborn Screening and Genetics Resource Center's "Performance Evaluation and Assessment Scheme." The program will work to enhance the disaster preparedness/contingency plan. Also there are plans to participate in Region IV's (Great Lakes Region) long-term follow-up data for MS/MS screened conditions upon IRB (Institutional Review Board) approval and with patient consent. Collection and analysis of long-term follow-up data can inform the system on the effectiveness of newborn screening, successful strategies (including best practice for medical treatment for positive outcomes), and provide a conduit of communication to assure continuity of services for children with special health care needs due to metabolic or other screened conditions. Nebraska's participation in this long-term data collection system will be made possible through funding from the Heartland Regional NBS and Genetics Collaborative.

CONTINUING ACTIVITIES

Education: Educational activities by the NNSP will continue via publication of the Annual Report, and as needed through hospital and physician mailings. Opportunities for on-site education are always available upon request from hospitals.

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Laboratory Testing: The contract with PerkinElmer Genetics screening laboratory is a one-year contract beginning July of 2008, renewable for four additional years. New for the FY 09 contracts are expanded areas receiving Saturday specimen pick-up service. The laboratory and program will continue to collaborate on improved efficiencies and quality throughout the system.

Follow-up, Tracking and Referral: Nebraska is very fortunate to have two highly skilled and dedicated follow-up staff, Krystal Baumert and Karen Eveans. They will continue to track every newborn to be sure they received an appropriate screen. Those reported as transferred, drawn early, transfused, unsatisfactory, inconclusive and presumptive positive specimens will be followed until complete. In addition, they will continue to facilitate confirmatory testing and referral for diagnostic and treatment services. Ongoing and annual review and updating of short-term follow-up procedures will be completed.

Confirmatory Testing: The program will continue to work with specialists and the Newborn Screening Advisory Committee to ensure procedures recommended for confirmatory testing are communicated effectively to practitioners. The program will continue to use the ACMG ACT (Action Sheets) for all positive results from tandem mass spectrometry to help physicians in Nebraska know what "next steps" to take when faced with a positive screening result for any of these rare conditions. The program will use other ACT sheets specific to Nebraska's screen and resources for all other screened conditions.

Diagnosis: Practitioners are strongly urged to consult with the pediatric specialist appropriate to the condition for which a newborn has a positive or abnormal screening result. The program will help link the newborn's primary care provider with specialists, when needed.

Treatment: Access to treatment will continue to be an issue the program will monitor. The statutorily required payment for metabolic foods and formula will continue. The program will continue to monitor the issues associated with access to treatment and seek ways to ensure funding is sufficient to meet affected individuals' needs.

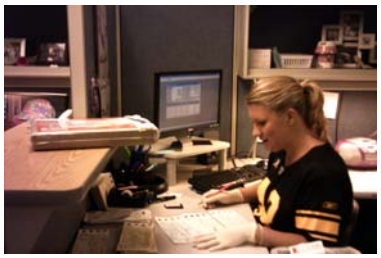
Quality Assurance Monitoring: The Program and Advisory Committee will continue to review and act on quarterly quality assurance plan data as well as respond to trends identified with any problems in the interim periods. Measures included in the quarterly QA reports will be expanded and "Quality Improvement Hints" publications will continue to be sent to individual hospitals for their own evaluation and comparison with statewide numbers.

FACES and NAMES

STATE NEWBORN SCREENING PROGRAM STAFF

(Left to Right)
Mike Rooney, Administrative Assistant
(Metabolic Foods, Translation & Distribution of Parent Education Brochures)
Karen Eveans, Follow-up Specialist
(Hemoglobinopathies, CF, Unsatisfactory Specimens & Drawn Early Specimens)
Krystal Baumert, Follow-up Coordinator
(Metabolic and Endocrine Conditions, Transfusions, Data Systems, Monthly Match and Home Births)
Julie Miller, Program Manager
(Regulations, Contracts, Policy/Procedure, Quality Assurance, Parent & Professional Education)

PERKIN ELMER GENETICS SCREENING LABORATORY STAFF



Accessioning

Client Services Team

Data Processing

Tandem Mass Spectrometry team

Laboratory

DNA Lab

Laboratory

Genetic Counselors

Information Technology Team

John Sherwin, PhD
Perkin Elmer Genetics
Director of Laboratory Operations
Perkin Elmer Genetics

Bill Slimak
Vice President/General Manager
Newborn Screening Laboratory
Perkin Elmer Genetics

Joseph Quashnock, PhD
Laboratory Director
Newborn Screening Laboratory
Perkin Elmer Genetics

Dennis Freer, PhD
Consultant, Biochemistry
Perkin Elmer Genetics

Jim DiPerna, PhD
Manager of Operations
Tandem Mass Spectrometry
Perkin Elmer Genetics

Zhili Lin, PhD
Director
Research and Development
Perkin Elmer Genetics

Bob Wise
Sr. Manager of Operations
(Biochem, DNA, Client Svs., Spec Processing)
Perkin Elmer Genetics

EARLY HEARING DETECTION AND INTERVENTION (NEWBORN HEARING SCREENING)

What is Newborn Hearing Screening?

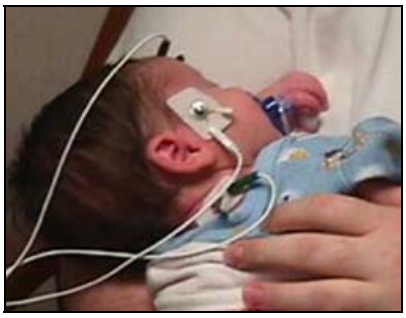
Significant hearing loss is one of the most common birth conditions, with an estimated incidence rate of one to three per thousand live births. Before newborn hearing screening, many hearing losses were not diagnosed until 2 ½ to 3 years of age. Left undetected, hearing loss in infants can negatively impact speech and language acquisition, academic achievement, and social and emotional development. If detected early, the negative impacts can be diminished and even eliminated through early intervention.

Newborn hearing screening is an essential preventive public health program. It meets the following prerequisites for a population screening program:

- Condition is sufficiently frequent in the screened population
- Condition is serious or fatal without intervention
- Condition must be treatable or preventable
- Effective follow-up program is possible

In 2000, the Infant Hearing Act established newborn hearing screening in Nebraska. The statute required birthing facilities to educate parents about newborn hearing screening, to include hearing screening as part of the standard of care and to establish a mechanism for compliance review by December 2003. The act also required that regulations be promulgated to mandate newborn hearing screening if less than 95% of newborns in the state received a hearing screening. This report presents the status of newborn hearing screening in Nebraska during 2007 (see Nebraska Early Hearing Detection and Intervention Data for 2007).

Newborn hearing screening requires objective physiologic measures to detect hearing loss in newborns and young infants. There are two basic techniques that birthing facilities in Nebraska use to screen newborns for hearing loss. Both are easily recorded in newborns and are non-invasive measures of physiologic activity that underlie normal auditory functioning.



The most frequently used screening technique is measurement of otoacoustic emissions, or OAEs. A miniature earphone and microphone are placed in the newborn's ear canal, low intensity sounds are presented, and responses produced by the inner ear are measured. The second screening technique, Auditory Brainstem Response, or ABR, uses small electrodes to detect certain brainwaves in response to sounds that are presented by a miniature earphone. For both methods, the response of each ear is measured. OAE and ABR are both reliable and accurate. Screening can occur as early as 12 hours of age, preferably with the newborn sleeping, and averages from five to 20 minutes to complete.

If a response is not detected for one or both ears, the result is a "refer" (did not pass). A "refer" to the screening test indicates that a hearing loss *may* exist but there are also other factors that may have contributed. A "refer" does indicate that a second screening is necessary to determine if the other factors, such as vernix in the ear canal, fluid in the middle ear cavity, movement, equipment failures, or inexperience of the tester, contributed to the initial result. A "refer" on the second screening indicates the need for further testing to confirm or rule out a hearing loss and, if hearing loss is present, to begin to identify the type and degree of the loss. Each birthing facility has established a newborn hearing screening protocol that identifies how the screening will be administered, the recording and reporting procedures, how refers will be handled, i.e., re-screen as an inpatient with the same or different screening technique or re-screen as an outpatient, and quality assurance measures.

EARLY HEARING DETECTION AND INTERVENTION SYSTEM

System Elements

The Nebraska Early Hearing Detection and Intervention (NE-EHDI) Program strives to fulfill the four 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; and
- 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.

Newborn hearing screening is one aspect of a comprehensive, integrated Early Hearing Detection and Intervention (EHDI) system. The Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs (Joint Committee on Infant Hearing, 2007) identifies eight principles that outline the elements of an EHDI system. The eight principles are:

1. All infants should have access to hearing screening using a physiologic measure no later than one month of age.
2. All infants who do not pass the initial hearing screening and the subsequent rescreening should have appropriate audiologic and medical evaluations to confirm the presence of hearing loss no later than three months of age.
3. All infants with confirmed permanent hearing loss should receive early intervention services

as soon as possible after diagnosis but no later than six months of age. A simplified, single point of entry into an intervention system appropriate for children with hearing loss is optimal.

4. The EHDI system should be family-centered, with infant and family rights and privacy guaranteed through informed choice, shared decision-making, and parental consent in accordance with state and federal guidelines. Families should have access to information about all intervention and treatment options and counseling regarding hearing loss.
5. The child and family should have immediate access to high-quality technology, including hearing aids, cochlear implants and other assistive devices when appropriate.
6. All infants and children should be monitored for hearing loss in the medical home. Continued assessment of communication development should be provided by appropriate professionals to all children with or without risk indicators for hearing loss.
7. Appropriate interdisciplinary intervention programs for infants with hearing loss and their families should be provided by professionals knowledgeable about childhood hearing loss. Intervention programs should recognize and build on strengths, informed choices, traditions, and cultural beliefs of the families.
8. Information systems should be designed and implemented to interface with electronic health records and should be used to measure outcomes and report the effectiveness of EHDI services at the patient, practice, community, state, and federal levels.

The screening, referral and audiological evaluation protocols developed by the Advisory Committee of the Nebraska Newborn Hearing Screening Program, as it was known in 2001, are consistent with the first three principles of the Joint Committee on Infant Hearing (JCIH). The protocols are for hearing screening to be completed during birth admission, audiological diagnostic evaluation to begin prior to six weeks of age to minimize the need for sedation, and appropriate early intervention activities to be initiated by six months of age. The NE-EHDI Program continues to develop new approaches to create a comprehensive EHDI system as outlined in the JCIH principles.

The early hearing detection and intervention system in Nebraska is composed of five functional elements: hearing screening at birth, confirmatory testing, medical evaluation, early intervention, and tracking and surveillance. One or more groups of professionals in a variety of settings assume responsibility for each element of the system. An overview of each of the elements and the primary activities are presented below. Included in this discussion are the Nebraska Revised Statute citations and the recommended protocols established by the Department of Health and Human Services through the Nebraska Early Hearing Detection and Intervention Advisory Committee.

Hearing Screening at Birth

Birthing facilities in Nebraska have five primary activities related to screening the hearing of newborns:

1. The parent(s) of newborns are educated about the hearing screening, the likelihood of hearing loss in newborns, the importance of follow-up, community resources (including early intervention services), and normal auditory, speech and language development (Neb. Rev. Stat. §71-4740). If risk

factors are present, hospital personnel educate parents to evaluate hearing every six months. *Note:* The Department of Health and Human Services is responsible for educating the parent(s) for newborns not born in a birthing facility (Neb. Rev. Stat. §71-4740).
2. A hearing screening test is part of each birthing facility's standard of care for newborns, effective 12/1/03 (Neb. Rev. Stat. §71-4742). Following hospital protocol for the procedure, each newborn's hearing in each ear is screened during birth admission using OAE and/or ABR screening techniques.
3. A mechanism for compliance review is established for each birthing facility (Neb. Rev. Stat. §71-4742).
4. Results of the hearing screening for each newborn are reported electronically to the NE-EHDI Program and should be reported to the newborn's primary care provider.
5. Annual reports are calculated based on the electronic reports for each occurrent birth and indicate the numbers of babies born in the birthing facility, recommended for screening, received screening during birth admission, passed screening, did not pass screening, and recommended for monitoring and follow-up (Neb. Rev. Stat. §71-4739).

Confirmatory Testing

Newborns who have referred for one or both ears on the inpatient hearing screening should receive an outpatient screening or an audiological diagnostic evaluation to confirm the presence of a hearing loss and to determine the type and degree of the hearing loss. The primary recommended activities that comprise the confirmatory testing component are:

1. An outpatient screening may be conducted within one to three weeks if the baby "refers" on the first screening. The outpatient screening for those that "refer" during birth admission may occur at the birthing facility or at a confirmatory testing facility.
2. If the infant "refers" on the outpatient screening, the testing should proceed immediately to a comprehensive diagnostic evaluation. This evaluation minimally includes measures of middle ear function (high frequency tympanometry), auditory sensitivity (air- and bone-conducted ABR), confirmatory measures (parent observations) and, depending upon the developmental age, behavioral audiological assessment (Visual Reinforcement Audiometry). Other measures may be included, as indicated.
3. Depending upon a variety of factors, referrals are made for further evaluation, diagnosis, treatment and services. These referrals may be made to medical specialists and/or early intervention services.
4. Results of the initial and comprehensive audiological diagnostic evaluation are provided to the primary care physician and NE-EHDI Program.
5. Annual reports are submitted to the NE-EHDI Program that indicate the number of newborns who return for follow-up testing, the number who do not have a hearing loss, and the number who do have a hearing loss (Neb. Rev. Stat. §71-4739).

Medical Evaluation

The infant's primary care provider (PCP) has the key role in the follow-up for those who "refer" on the initial hearing screening during the birth admission. Building on the concept of a medical home (*Guidelines for Pediatric Medical Home Providers*, American Academy of Pediatrics), the PCP has the primary role in identifying and accessing the medical and non-medical services needed to help children and their families achieve their maximum potential. The primary activities that comprise the medical element of the newborn hearing screening system are:

1. Birthing hospital notifies PCP of the newborn's hearing screening results.
2. PCP or designee per hospital procedure informs parents of hearing screening results and need for re-screening.
3. NE-EHDI Program notifies PCP about the hearing screening status and need for follow-up evaluation for those who did not pass the inpatient hearing screening or were discharged without a screening.
4. PCP (or staff), hospital, or test provider schedules an appointment for an outpatient screening to be completed in one to three weeks and notifies parents.
5. Provider of outpatient screening notifies PCP of results.
6. PCP notifies NE-EHDI Program of outpatient hearing screening results.
7. If "refer," PCP makes referral for comprehensive diagnostic evaluation, educates parents about need for evaluation, and makes referral to early intervention services.
8. If hearing loss is confirmed, PCP or diagnostic evaluator refers newborn/infant for complete medical and/or neuro-sensory evaluation and early intervention Services.

Early Intervention

Early intervention is an individualized program of services and supports based on the needs of the individual and family. Part C of the Individuals with Disabilities Education Act (IDEA) authorizes the creation of early intervention programs for infants and toddlers with disabilities. In Nebraska, the Early Development Network (EDN) provides services coordination for eligible families to identify and link with needed services and to work with multiple providers to ensure that services are provided. The recommended practices for the primary early intervention activities within the EHDI system are:

1. Upon receiving a referral, the EDN services coordinator immediately contacts the appropriate school district to begin the MDT process.
2. The services coordinator contacts the parent(s)/guardians to explain the importance of having a teacher of the deaf involved early and to obtain support for an initial joint meeting with the family.
3. Upon receiving verbal permission from the parent, the services coordinator contacts the Regional Programs for Students Who are Deaf or Hard of Hearing. The Regional Program

coordinator contacts the school district to determine the appropriate teacher of the deaf to attend the joint meeting with the family.

4. The NE-EHDI Program is included on the Authorization for Release of Information form.
5. If the family would like support from organizations for young children with hearing loss and their families, the following organizations may be included on the "Release of Information" form to allow the parents' contact information to be shared: Regional Programs for Students Who are Deaf or Hard of Hearing, PTI-NE, and/or Hands and Voices.
6. A NE-EHDI Health Portfolio/Resource Guide for parents is provided.

Tracking and Surveillance

The NE-EHDI Program has developed, based on the requirements identified in the Infant Hearing Act (Neb. Rev. Stat. §71-4735 - §71-4744) and the NE-EHDI Advisory Committee's recommended protocols. The requirements are to "…determine and implement the most appropriate system…to track newborns and infants identified with a hearing loss" and "…to effectively plan and establish a comprehensive system of developmentally appropriate services for newborns and infants who have a potential hearing loss or who have been found to have a hearing loss and shall reduce the likelihood of associated disabling conditions" (Neb. Rev. Stat. §71-4737). Activities of the NE-EHDI Program include:

1. Develop, implement, and monitor statewide systems to track newborns with or at risk of hearing loss (Neb. Rev. Stat. §71-4737) and adopt and promulgate rules and regulations to implement the Infant Hearing Act (Neb. Rev. Stat. §71-4742 and §71-4744).
2. Gather required data and generate annual reports (Neb. Rev. Stat. §71-4739 and §71-4741).
3. Establish guidelines for referral to early intervention services (Neb. Rev. Stat. §71-4743).
4. Educate parents with out-of-hospital births about newborn hearing screening (Neb. Rev. Stat. §71-4740)
5. Apply for all available federal funding to implement the Infant Hearing Act (Neb. Rev. Stat. §71-4738).

EARLY HEARING DETECTION AND INTERVENTION DATA FOR 2007

Newborn Hearing Screening Data Reported for 2007

Birthing Facility Screening Programs

The number of birthing facilities conducting newborn hearing screening has increased rapidly since 2000, when only 11 hospitals were conducting either targeted or universal newborn hearing screening. In 2007, 100% of the birthing facilities in Nebraska were conducting hearing

Birthing Facilities Conducting Newborn Hearing Screenings (2000-2007)

Year	Number of Birthing Facilities in Nebraska	Number Conducting Newborn Hearing Screening	Percentage Conducting Newborn Hearing Screening
2000	69	11	16%
2001	69	24	35%
2002	69	57	83%
2003	67	67	100%
2004	67	67	100%
2005	65	65	100%
2006	63	63	100%
2007	63	63	100%

Table 1

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. Sixty-two (62) of the 63 birthing hospitals conducted the hearing screening during the birth admission and one conducted the screening on an outpatient basis following discharge.

Annual Birthing Facility Reports

Birthing facilities are required to annually report specific information about their newborn hearing screening programs to the Department of Health and Human Services (Neb. Rev. Stat. §71-4739). The new ERS-II data system, an integrated module of the state's Vital Records system, automatically calculates these figures for each birthing facility.

Birthing Facility Reports of Required Aggregate Data (2007)

Number of newborns born (includes out-of-hospital births with birth certificates)	27,117
Number of newborns and infants recommended for a hearing screening test	26,042
Number of newborns who received a hearing screening during birth admission	26,737
Number of newborns who passed a hearing screening during birth admission	25,753*
Number of newborns who did not pass a hearing screening during birth admission	984*
Number of newborns recommended for monitoring, intervention, follow-up care	645*

*Figures include hearing screenings for babies transferred to Children's Hospital, a non-birthing facility

Table 2

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 ten languages. Birthing facilities reported educating almost all parents (25,965 or 95.7%) about newborn hearing screening, hearing loss and normal speech and language development in 2007. Neb. Rev. Stat. §71-4740 requires the 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. Parent education material was sent to the parents of the 77 babies who were not born in a hospital.

Newborns Receiving a Hearing Screening

The Infant Hearing Act requires that rules and regulations be adopted and promulgated if the annual percentage rate of newborns who receive a hearing screening during birth admission is less than 95% by December 1, 2003, or at any time thereafter. Hearing screening results reported for occurrent births in 2007 show that 98.6% of the 27,117 births were screened during birth admission or prior to discharge to home. The numbers of newborns screened during birth admission increased dramatically since reporting began in 2000, when only slightly more than one third of newborns received a hearing screening during birth admission (see Table 3). This increase in the numbers of newborns receiving a hearing screening corresponds to the increase in the number of hospitals adopting newborn hearing screening as the standard of care for newborns and the support of sub-grants through the Nebraska Health Care Cash Fund to purchase screening equipment in 2002 and 2003.

Newborns Receiving a Hearing Screening Prior to Discharge to Home (2000-2007)

	2000	2001	2002	2003	2004	2005	2006	2007
Number Receiving a	8,978	15,272	22,615	25,275	25,966	26,179	26,615	26,737
Percent Receiving a	36%	61%	89%	97%	98%	99%	99%	99%

Table 3

Newborns Discharged Without a Hearing Screening

During 2007, the ERS-II reports available for each birthing facility indicated that there were 337 newborns who did not receive a hearing screening during birth admission because the newborn

expired prior to screening (143), was discharged to home prior to screening (177) or the parent refused (7). Ten records had insufficient data to determine the results.

Birth Admission Refer Rates

The ERS-II reports available for each birthing facility indicated that 984 newborns did not pass ("refer") the hearing screening during birth admission, or prior to discharge to home for those babies who were transferred to another hospital. Of the newborns with hearing screenings conducted during the birth admission, the refer rate for all birthing facilities was 3.7% during 2007, which compares favorably with refer rates for previous years at Nebraska birthing facilities (see Table 4).

Birth Admission Refer Rates (2002-2007)

	2002	2003	2004	2005	2006	2007
Refer rate for all birthing facilities	3.7%	3.6%	3.5%	3.4%	3.8%	3.7%

Table 4

As discussed previously in this report, there are two measurement techniques used to conduct newborn hearing screening: Otoacoustic Emissions (OAE) and Auditory Brainstem Response (ABR). Almost half of the birthing hospitals in Nebraska are using OAE-only, almost one third are using ABR-only, and the remaining birthing hospitals are using a 2-step method (OAE, followed by ABR if the initial screening is a "refer"). The "refer" rates differ for the three techniques with the OAE-only having the highest refer rate (see Table 5).

Refer Rates for Hearing Screening Techniques (2007)

	OAE-only	ABR-only	2-Step
Number of Birthing Facilities	32	21*	10
Refer Rate	10.5%	2.1%	2.5%

*includes Children's Hospital

Table 5

Monitoring, Intervention and Follow-up

Another ERS-II report available for each birthing facility is the number of newborns recommended for monitoring, intervention, and follow-up care. In 2007, 645 (65.5% of the babies who referred) were recommended for monitoring, intervention and follow-up care by the birthing facilities. The results of previous years are: 709 (85% of refers) in 2002, 676 (74% of refers) in 2003, 793 (86% of refers) in 2004, 863 (95% of refers) in 2005, and 961 (96% of refers) in 2006. The percentage for 2007 is lower than in previous years because the new electronic reporting system included a specific data field that needed to be checked for each individual report. Several large birthing facilities did not consistently check that data field, resulting in a lower number of recommendations being reported. Regardless of whether the field was checked or not, the NE-EHDI Program's tracking and follow-up processes were followed for each baby who did not pass the hearing screening during birth admission.

The NE-EHDI Program also tracked 1,417 newborns who were transferred to neonatal intensive care units or to hospitals with a higher level of care in Nebraska and surrounding states prior to receiving a hearing screening.

Out-of-Hospital Births

Although parent education was provided to the parents of all reported out-of-hospital births during 2007, only 44.2% (34 of 77) of out-of-hospital births were screened (see Table 6) and 43 were not screened or the results were not submitted to NE-EHDI Program.

Out-of-Hospital Births (2001 – 2007)

	2001	2002	2003	2004	2005	2006	2007
Out-of-hospital births	93	99	70	60	55	68	77
Number screened	5	16	12	13	15	30	34
Percentage screened	5%	16%	17%	22%	27%	44%	44%

Table 6

Confirmatory Testing/Audiologic Data Reported for 2007

The Advisory Committee for the NE-EHDI Program identified the initial level of the follow-up hearing test for many newborns as an outpatient screening of the newborn's hearing. For those newborns and infants who pass this initial level of follow-up, no further audiological evaluation would be needed, unless there are risk factors present that would warrant periodic monitoring.

Since the majority of newborns will pass this second screening, considerable cost savings can result by using either the OAE and/or ABR screening technique rather than proceeding directly to a complete diagnostic audiological evaluation. The Advisory Committee's Audiological Diagnostic Protocol recommends that the screening facility should be prepared to provide comprehensive audiological diagnostic procedures if the outpatient re-screening results indicate a "refer" status. However, many communities that do not have pediatric audiology services readily available have opted to have the second screening occur at the birthing facility on an outpatient basis.

Annual Confirmatory Testing Facility Reports

Neb. Rev. Stat. §71-4739 requires confirmatory testing facilities to report the following:

- Number of newborns and infants who return for a follow-up hearing test
- Number of newborns and infants who do not have a hearing loss based upon the follow-up hearing test
- Newborns and infants who are shown to have a hearing loss based upon the follow-up hearing test

Each year, data regarding the follow-up hearing tests at confirmatory testing (audiologic evaluation) facilities have been gathered by surveying the audiologists in Nebraska. Twenty-

eight (28) of 33 confirmatory testing facilities responded in 2007, representing 70 licensed audiologists. The results of those surveys are included in Table 7.

Required Follow-up Hearing Test Data Reported by Audiologists

	Re-screenings	Diagnostic Evaluations
Number of newborns/infants receiving a follow-up hearing test	706	174
Number of newborns/infants without a hearing loss	543	64
Number of newborns/infants with a hearing loss	163 ("refer")	110

Table 7

Diagnosis of Hearing Loss

The number of infants diagnosed with a hearing loss in Nebraska is reported in two ways: 1) aggregate reports submitted by audiologists with the number of infants shown to have a hearing loss based on follow-up tests (required by Neb. Rev. Stat. §71-4739) and 2) the individual diagnostic reports submitted to NE-EHDI Program by audiologists or primary care providers. Aggregate reports may include duplicate entries. Statutory authority to require audiologists to report on all newborns and infants who receive audiological evaluations does not exist, so a one-to-one correspondence between the individual results reported to NE-EHDI Program and the required annual aggregate reporting does not exist. As shown in Table 7, the aggregate reports indicated that 110 infants were identified with either transient or permanent hearing loss.

Type and Degree of Hearing Loss

Analysis of the aggregate confirmatory testing reports submitted to the NE-EHDI Program indicates that 55 of the infants with hearing loss meet the criteria for a permanent hearing loss (PHL). Forty four (44) of the infants with permanent hearing loss were identified with a bilateral hearing loss, 39% in the mild to moderate range and 61% in the severe to profound range. The remaining 11 infants were identified with a unilateral hearing loss, 73% of which were in the mild to moderate range. The aggregate reports indicated that only 28 infants had been fit with amplification.

Type and Degree of *Permanent* Hearing Loss, 2007 (n = 55)

Degree ► / Type ▼	Bilateral Mild–Moderate	Bilateral Severe–Profound	Unilateral Mild–Moderate	Unilateral Severe-Profound
Sensorineural	17	24	3	3
Conductive	0	-	1	-
Undetermined	0	3	4	0

Table 8

The estimates of the incidence of permanent hearing loss in newborns range between 1 to 3 per thousand births nationally. Based on the birth rate in Nebraska during 2007 (27,117), an

estimated 27 to 81 newborns would be identified with PHL. The incidence of PHL in Nebraska for babies born in 2007 reported in aggregate reports is 2 per thousand newborns.

Tracking and Follow-up Results for 2007

The NE-EHDI Program tracked the 1,161 newborns who were reported as not passing a newborn hearing screening during birth admission. Of those, 984 newborns referred on the birth admission hearing screening and 177 newborns were discharged to home prior to receiving a hearing screening. These were the newborns who were tracked through follow-up outpatient screenings, diagnostic evaluations and early intervention services.

Rate of Follow-up Outpatient Screening and Confirmatory Testing

Follow-up services include outpatient hearing screenings, audiologic diagnostic evaluations, or a combination of the two, depending upon clinical findings. Hearing screenings were conducted at some birthing facilities or at confirmatory testing (audiology) facilities. Outpatient hearing screenings were provided by 53 birthing facilities for 509 newborns, of which 380 were re-screenings due to a "refer" on the birth admission screening and 129 were initial hearing screenings for newborns who had been discharged to home prior to receiving a hearing screening. The aggregate reports from the confirmatory testing facilities indicated that 706 newborns received screenings and 174 received audiologic diagnostic evaluations. With aggregate reporting, it is not possible to determine an unduplicated count, since some infants, especially those with middle ear dysfunction and an accompanying transient conductive hearing loss, may be screened or evaluated multiple times at one or more facilities.

Follow-up Services and Outcomes

Based on individual reports submitted to NE-EHDI, follow-up screening and/or diagnostic evaluations were completed for 1,010 infants, with 960 having normal hearing and 50 being identified with a permanent hearing loss. The evaluation process is still in progress for 12 infants, the parents of 2 infants refused to complete the follow-up, and 6 infants expired before completion of the follow-up services. There were 131 newborns needing follow-up for whom follow-up services were not initiated, were initiated but not completed, or were not reported to NE-EHDI Program.

Individual reports were received by the NE-EHDI Program for 52 infants diagnosed with a PHL. Two of these were newborns who had passed the newborn hearing screening during birth admission. The average age at the first audiologic diagnostic evaluation for these infants was 91.1 days. The average age at confirmation of PHL was 121.7 days with slightly more than half (53.8% or 28 of 52 infants) having a hearing loss confirmed before 90 days of age which is the recommended guideline. Appendix A tracks the services and outcomes of the 1,161 newborns needing follow-up services through the EHDI system and indicates the results for those infants.

Timeliness of Follow-up Screening/Testing

To meet the state and national guidelines of "1-3-6" (hearing screening completed by 1 month,

audiological diagnostic evaluation initiated 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. For the newborns who received follow-up services, 74.7% received an outpatient screening or diagnostic evaluation prior to 1 month of age. The peak of follow-up activity occurred at approximately 2 weeks of age (see Chart 1). The average age of follow-up service initiation was 29.4 days and the average age at completion of follow-up 37.0 days.

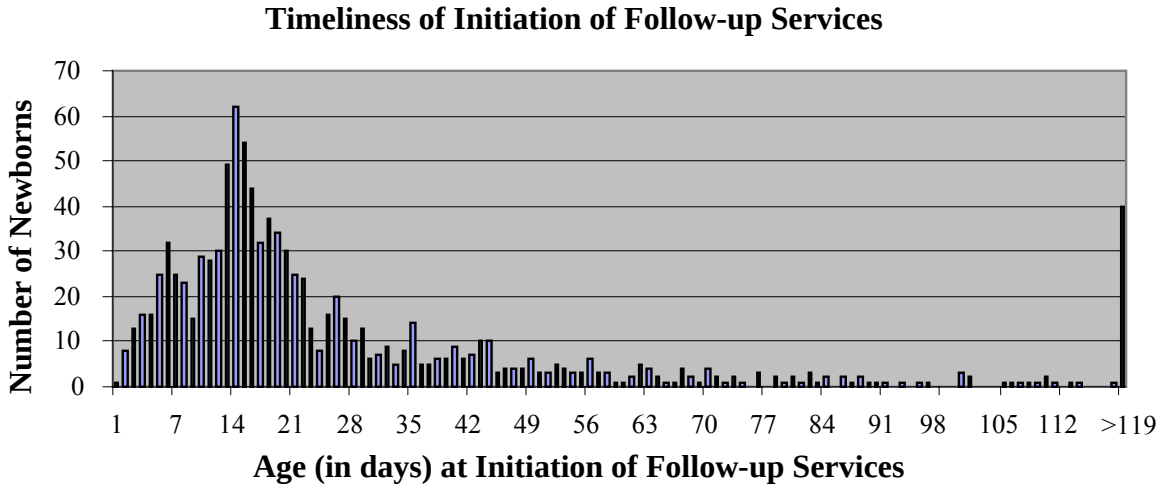


Chart 1

Undetermined Hearing Status

Neb. Rev. Stat. §71-4742 states: "…it is the goal of this state to achieve a one-hundred-percent screening rate." While Nebraska has made great strides in developing a comprehensive early hearing detection and intervention system, there are also infants for whom the status of their hearing is not known. In 2007, there were 354 newborns whose hearing status has not been objectively established:

- 98 infants had no outpatient follow-up initiated or reported.
- 33 newborns had a follow-up re-screening or diagnostic evaluation that was inconclusive, with no additional follow-up received or reported.
- 12 were identified with hearing problems associated with middle ear dysfunction, but additional follow-up evaluations have not yet been completed.
- 43 of the out-of-hospital births were not screened or the results were not submitted to NE-EHDI Program.
- 7 parents refused the hearing screening during birth admission.
- 2 parents refused the hearing screening after follow-up was initiated, but before it was completed.
- 143 newborns expired prior to receiving an inpatient hearing screening.
- 6 expired before outpatient hearing services were initiated or completed.
- 10 records had insufficient data to determine the results.

Based on the analysis of the hearing screening and follow-up records, the hearing status (normal hearing or permanent hearing loss) of 98.7% of the 27,117 newborns was confirmed.

Early Intervention

The purpose of the Infant Hearing Act (Neb. Rev. Stat. §71-4735) is to "obtain needed multidisciplinary evaluation, treatmen, 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." Records for the Early Development Network, Nebraska's Part C Early Intervention Program, indicate that 40 (76.9%) of the 52 infants with PHL were verified for special education services. Verification for 35 of the 40 infants was completed prior to 6 months of age and 5 were verified after 6 months of age. Seven (7) infants were not referred to EDN, 2 had insufficient data available after referral, 2 moved and the parents of one infant withdrew prior to verification. Thirty four (34) of the infants diagnosed with a PHL have a medical home.

Summary

- All the current birthing hospitals in Nebraska were conducting newborn hearing screening in 2007. All but one had conducted the hearing screenings during the birth admission.
- In 2007, birthing hospitals reported screening the hearing of 98.6% of newborns during birth admission or prior to discharge to home for those babies who were transferred to another hospital. The benchmark of 95% of newborns having a hearing screening during birth admission, established by Neb. Rev. Stat. §71-4742, has been met.
- The overall "refer" (did not pass) rate during 2007 for hearing screening during birth admission was 3.7%.
- In 2007, follow-up hearing screenings or audiologic evaluations were initiated within one month of birth for 74.7% of those newborns for whom follow-up activities were provided.
- The average age at the time of the initiation of follow-up hearing screening or diagnostic evaluation was 29.4 days and the average age at completion was 37.0 days.
- For the 52 infants identified with a permanent hearing loss, the average age at the first audiologic diagnostic evaluation was 91.1 days and the average age at confirmation of hearing loss was 121.7 days.
- There were 354 infants whose hearing was not objectively established. Of those, 149 expired before receiving or completing hearing screenings.
- The incidence of permanent hearing loss identified and reported to NE-EHDI Program (2 per thousand in 2007) appears to be within the anticipated range of 1 to 3 per thousand.
- Over 75% of the infants with a permanent hearing loss were verified for early intervention services.

ACTIVITIES

Funding

Health Resources Services Administration/Maternal and Child Health Bureau

The NE-EHDI Program received grant funds from the Health Resources Services Administration/Maternal and Child Health Bureau (HRSA/MCHB) to fund the basic operations of the NE-EHDI Program for the third year of a three-year funding cycle. Funding for the 12-month period from 4/1/07 to 3/31/08 was $125,000, with an additional $41,002 available from funds carried over from the previous year. The NE-EHDI Program goals in 2007 for the HRSA/MCHB grant were:

System Goal 1 – The hearing of all newborns in Nebraska will be screened during the birth admission or, if born out-of-hospital, by one month of age.

System Goal 2 – All newborns who "refer" on the initial outpatient hearing re-screening will complete an audiologic diagnostic evaluation prior to 3 months of age.

System Goal 3 – All infants with confirmed hearing loss will begin receiving early intervention services prior to six months of age.

System Goal 4 – All infants with a confirmed hearing loss will have a medical home.

System Goal 5 – Families of young children with a confirmed hearing loss will have access to a family-to-family support system.

System Goal 6 – The hearing of young children in Nebraska will be screened at various times prior to age 3.

System Goal 7 – Hearing health professionals will increase their capacity to provide appropriate services to young children.

System Goal 8 – NE-EHDI Program will provide an effective structure for the newborn hearing screening and intervention system in Nebraska.

Centers for Disease Control and Prevention

The NE-EHDI Program received funding from the Centers for Disease Control and Prevention for the third year of a three year Early Hearing Detection and Intervention Tracking, Surveillance and Integration cooperative agreement. Funding for the 12-month period from 7/1/07 to 6/30/08 was $145,850, with an additional $29,876 available from funds carried over from the previous year. The 2007 goals of the cooperative agreement were:

Goal 1 – NE-EHDI Program will have hearing screening results for all occurrent births in Nebraska.

Goal 2 – Pediatric audiologic evaluations and risk factors will be electronically reported to NE-EHDI Program.

Goal 3 – The NE-EHDI Program data system, integrated with electronic birth certificate registry, will be electronically linked with related child data systems.

Goal 4 – A formative and summative evaluation of the NE-EHDI Program tracking, surveillance and integration project will be conducted and the results disseminated.

Title V/Maternal and Child Health Block Grant

The NE-EHDI Program was approved to receive $62,500 for the third year of a three-year funding cycle through the Title V/Maternal and Child Health Block Grant. The anticipated outcomes for 2007 were:

Outcome Statement 1: The hearing of all newborns born in Nebraska will be screened during the birth admission (if born out-of-hospital, by one month of age) and, if needed, they will complete an audiological diagnostic evaluation prior to 3 months of age and begin receiving early intervention services prior to six months of age

Outcome Statement 2: All infants with a confirmed hearing loss will have a medical home and their families will have access to a family-to-family support system.

Outcome Statement 3: Hearing health professionals will increase their capacity to provide appropriate services to young children.

Outcome Statement 4: The structure of the Newborn Hearing Screening Program will support the hearing screening and intervention system in Nebraska.

Advisory Committee

The NE-EHDI Program was developed based on the requirements identified in the Infant Hearing Act of 2000 and the recommendations by the Advisory Committee. Specific tasks to be accomplished by the Advisory Committee are 1) to continue to increase the representation of stakeholders, 2) to review and, as necessary, revise the existing protocols to incorporate the electronic data system, 3) to develop new reporting, tracking and follow-up protocols to effectively link the NE-EHDI Program and the early intervention systems, 4) to increase the program's responsiveness to the expanding cultural and linguistic communities in the state, 5) to support the development of an effective professional development system, and 6) to guide the long-term planning and evaluation of the EHDI system in the state.

The Advisory Committee of the NE-EHDI Program consists of 23 members representing medical, audiology, parents, public health, family support, and education stakeholders (see Appendix A). The Advisory Committee met quarterly during 2007. The Committee received comprehensive updates of all aspects of the NE-EHDI program and provided the following guidance:

1. Continued discussion and input into procedures to retrieve the dried blood spots from the newborn screening laboratory to assist with the identification of the etiology of confirmed hearing loss.
2. Development of the parent education materials, including the "Learning About Hearing" checklist.
3. Approval of revisions to the recommended audiological assessment and reporting protocol.
4. Review and suggestions for a Memorandum of Agreement with Early Head Start/Head Start

programs.
5. Review and suggestions for a Guide By Your Side program, a parent-to-parent support program.
6. Long-term planning that included the review, revision and refinement of the NE-EHDI Program mission statement, logic model, system goals, and program objectives and activities.

The Audiology Sub-committee finished revision of the infant audiologic assessment recommendations and the screening and diagnostic reporting form and process.

The Evaluation Sub-committee reviewed and provided input for revision and improvement of the NE-EHDI evaluation questions and plan, quality assurance reports for birthing facilities, implementation and further development of the integrated electronic data system, progress in implementing the CDC cooperative agreement, and dissemination activities.

The Family Support Work Group of the NE-EHDI Advisory Committee was established in 2007 to recommend and develop approaches to increase the parent-to-parent support available to families of young children identified with hearing loss.

Projects for 2007

Electronic Data System
The hearing screening module of the Vital Records Electronic Reporting System (ERS-II), developed by Netsmart Technologies, Inc., was implemented on January 1, 2007. The integrated reporting system, funded through the cooperative agreement with CDC, is based on the birth records and provides for the reporting of hearing screening results for all occurrent births in Nebraska. All birthing facilities were granted access to the hearing screening module and selected staff was trained to create records and report hearing screening results from December, 2006, to March, 2007. Ongoing training and technical assistance was provided as requested to birthing facility staff. A variety of administrative reports, including exception reports identifying overdue results, were developed by the NE-EHDI business analyst. Specifications were developed and cost quotes received from Netsmart Technologies, Inc., to revise the basic reporting module and the reports available, as well as expansion of the data system to include audiological results and birth defects.

Periodic Early Childhood Hearing Screening
To begin the process of implementing periodic early childhood hearing screening in Nebraska, the Early Childhood Hearing Outreach (ECHO) project, developed by National Center for Hearing Assessment and Management (NCHAM) and funded by the Office of Head Start, provided training and screening equipment in 2004 through 2006. During 2007 summary reports for the six Early Head Start programs that were conducting OAE hearing screenings were provided by NCHAM. Discussions and technical assistance by phone and in-person helped overcome difficulties in following-up with re-screenings and in timely submission of paperwork. Two health care clinics in Omaha, Boys Town Pediatric Primary Care and Fred LeRoy Health and Wellness Clinic, were trained to conduct OAE hearing screenings using NCHAM's Hear and Now training curriculum.

Family-to-Family Support
The NE-EHDI Program provided resources to support a statewide teleconference to organize a chapter of Hands and Voices, a national family-to-family support organization. Representatives from the Regional Program for Students Who are Deaf and Hard of Hearing, PTI-NE, Early Development Network, Nebraska Chapter Hands and Voices and NE-EHDI Program attended the 2nd Annual "Investing in Family Support" conference in Nashville. The workgroup updated the "Committing to Family Support" work plan to collaboratively strengthen the formal and informal types of family support in the state.

Together for Kids and Families
Nebraska's State Early Childhood Comprehensive Systems (SECCS) grant program Together for Kids and Families seeks to achieve optimum outcomes for Nebraska's young children and their families through comprehensive system planning and collaborative effort among stakeholders. The NE-EHDI Program Manager served on workgroups to implement the following strategies:
1 Develop and implement a collaborative initiative to promote the medical home approach as a standard of care for all children;
2 Establish a comprehensive program to promote regular recommended pediatric visits for children, following the American Academy of Pediatrics and Bright Futures guidelines;
3 Integrate parent to parent peer support systems into existing and new programs and services for families;
4 Develop capacity for an early childhood data monitoring system through creation of an ECCS data agenda.

National Initiative for Children's Healthcare Quality (NICHQ) Learning Collaborative
Nebraska was one of eight states selected to participate in a Learning Collaborative, funded by the Health Resources Services Administration/Maternal and Child Health Bureau and developed by NICHQ. The purpose of the project was to reduce the number of babies who are lost to follow-up by developing strategies that have been found to be effective. Program activities during the Learning Collaborative, which concluded in July, 2007, included reduction of the literacy level of parent materials, development of a parent checklist/roadmap, inclusion of parent phone number, primary language, and primary health care provider name on hospital reports, development of a 1-page audiologic evaluation reporting form, and development of scripts to explain hearing screening results to parents. Partners in the NICHQ Learning Collaborative included the Nebraska Medical Center, Fred LeRoy Health and Wellness Clinic, Boys Town Pediatric Primary Care, Boys Town National Research Hospital, Early Development Network (Part C), Medically Handicapped Children's Program, Regional Program for Students Who are Deaf or Hard of Hearing, Medicaid/Primary Care Unit, Together for Kids and Families and the NE-EHDI Program.

Nebraska Children's Hearing Aid Loaner Bank
Based on a feasibility study conducted during 2006, the Nebraska Children's Hearing Aid Loaner Bank (NCHALB) was organized during 2007. The purpose of the NCHALB is to provide immediate access to amplification for children when identified with a permanent hearing loss for an initial period of six months. Partners in the NCHALB are the University of Nebraska

– Lincoln Barkley Center audiology department, the Nebraska Association for the Education of Young Children (NeAEYC) and the NE-EHDI Program. Contracts were signed to provide funding to NeAEYC for fiscal management and to UNL-Barkley Center for administration of the NCHALB. An organizing committee was formed and funding proposals, a brochure and the Web site (www.unl.edu/barkley/nchalb/index.shtml) were developed. One hearing aid manufacturer, GN ReSound, donated 30 hearing aids to the NCHALB.

Dried Blood Spot Retrieval
The feasibility of retrieving the newborn dried blood spot for identification of congenital cytomegalovirus (CMV), Connexin 26 and 30, mitochondrial, and Pendred syndrome continued to be evaluated and prioritized. Identification of these factors, which are risk factors for later-onset hearing loss, can also assist in establishing the etiology of a congenital hearing loss. A work group of the Newborn Screening (blood-spot) Advisory Committee recommended the implementation of procedures to encourage physicians to get the audiological evaluation and diagnosis completed before 90 days so that they could request that the dried blood spot be returned so it could be tested for genetic causes of hearing loss, or at a minimum, CMV for those infants diagnosed with sensorineural hearing loss. Educational materials were developed and mailed to physicians, physician assistants and nurse practitioners. Beginning in October 2007, materials for parent consent and physician retrieval of the dried blood spots for babies identified with sensorineural hearing loss before three months of age were included with the basic information provided to the primary health care provider.

Projects Planned for 2008

Electronic Data System
The ERS-II data system will be revised to improve functionality and to incorporate additional reports for the birthing facility users. The system will be expanded to include the recording of audiologic evaluations and birth defects related to hearing loss. The administrative tracking and follow-up system will be further developed.

Periodic Early Childhood Hearing Screening
Training and support to OAE hearing screening programs in Early Head Start programs and health care clinics will continue using materials developed by NCHAM. Adaptation of data systems to more systematically record childhood hearing screening data will be explored. A Memorandum of Agreement will be pursued with Early Head Start/Head Start programs.

Single Point of Entry
A multidisciplinary workgroup will finalize a strategy for a single point of entry to early intervention services for parents of newborns/infants identified with a PHL. The anticipated outcome of this work is that the parents will be able to access timely and appropriate early intervention services through a recognized point of entry that is knowledgeable about hearing loss, its effects on young children, and available resources.

Family-to-Family Support
The "Committing to Family Support" work plan will be fully implemented. The Family Support Work Group of the NE-EHDI Advisory Committee will provide input for this component of the

program. Partnership with the Nebraska chapter of Hands and Voices will continue, including exploration of establishing a Guide By Your Side program to provide parent-to-parent support when a young child is identified with a permanent hearing loss.

Nebraska Children's Hearing Aid Loaner Bank
The NCHALB began providing loaner hearing aids to young children in January, 2008. The partnership between UNL Barkley Center, NeAEYC and NE-EHDI will continue. The NE-EHDI program will provide funds to administer the NCHALB and to purchase loaner hearing aids. Requests for funding will continue to be sent to potential funders.

Dried Blood Spot Retrieval
Strategies to retrieve the newborn dried blood spot for identification of genetic causes of hearing loss will continue to be evaluated and prioritized.

Website and Promotion
A website for the NE-EHDI Program will be developed as part of the Lifespan Health Services website. A promotional plan for the NE-EHDI Program will also be developed and implemented.

(Left to Right)

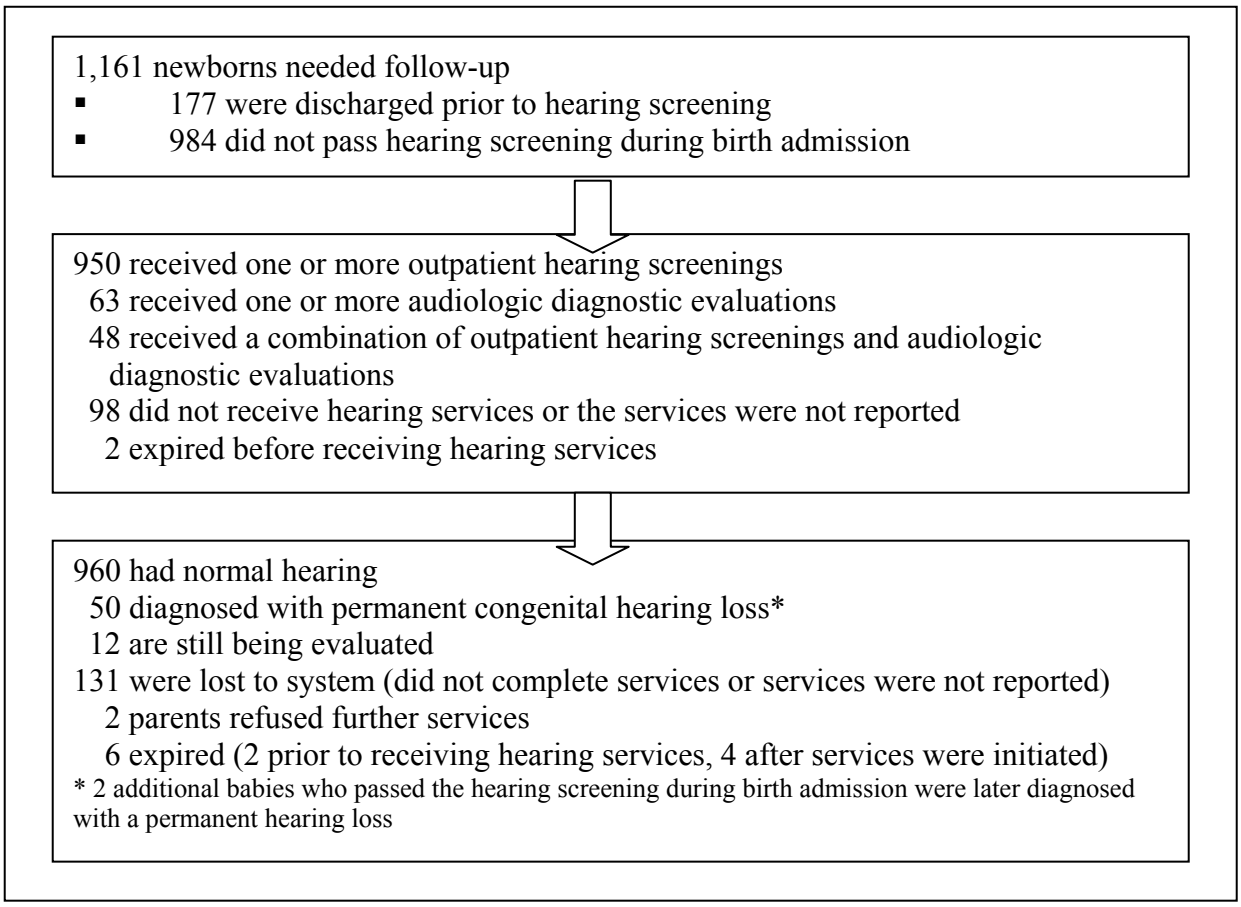
Jeff Hoffman, Program Manager
(Program planning, evaluation and management, systems development)

Kerry Julian, Staff Assistant
(Follow-up, patient education materials distribution, data management, special projects)

Jim Beavers, Business Analyst
(Data system planning and testing, development of reports, system security, training and technical assistance)

Appendix A.

Tracking of Follow-up Services and Outcomes



Appendix B.

NE-EHDI Advisory Committee Members

Committee Member	Group/Facility Represented
Steve Boney, PhD	Professor, Audiology University of Nebraska - Lincoln
Margaret A. Coleman	Consumer
Lora Langley	Public Health Nurse Ponca Tribe of Nebraska
Regina Watson	Hearing Screening Coordinator Tri County Area Hospital, Lexington
Kathy Beauchaine	Audiology Coordinator Boys Town National Research Hospital
Karen Rossi	Executive Director Omaha Hearing School
Stacie Ray	Parent, Clinical Practice Supervisor – Audiology University of Nebraska - Lincoln
G. Bradley Schaefer, M.D.	Geneticist Munroe-Meyer Institute, Nebraska Medical Center
Monica Seeland	Vice President Nebraska Hospital Association
Britt Thedinger, M.D.	Otologist Ear Specialists of Omaha
Donald M. Uzendoski, M.D.	Nebraska Chapter, American Academy of Pediatrics Early Hearing Detection and Intervention Chapter Champion
Robert Wergin, M.D.	Nebraska Academy of Family Physicians
Connie Shockley	Parent Support Coordinator Parent Training and Information-Nebraska
Eleanor Kirkland	Director Head Start State Collaboration Office (NDE)
Audrey Isaacson	Parent
Jennifer Johnson	Parent
Kenny Johnson	Parent
Rhonda Fleischer	Liaison Regional Programs for Deaf and Hard of Hearing Students (NDE)
Jeanne Garvin, M.D.	Medical Director Medically Handicapped Children's Program (CSHCN) (DHHS)
Charlie Lewis	Answers4Families
Amy Bunnell	Co-Lead Early Development Network (Part C) (DHHS)
Julie Miller	State Genetics Coordinator Newborn Screening Program (DHHS)
Krystal Baumert	Follow-up Coordinator Newborn Screening Program (DHHS)

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

Julie Miller, 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 Specialist (402) 471-6558
Hemoglobinopathies and Cystic Fibrosis, Drawn Early and Unsatisfactory Specimens
Mike Rooney, Administrative Assistant (NBS and EHDI) (402) 471-9731
Metabolic foods, translation and distribution of patient education materials

WEB PAGE: www.dhhs.ne.gov/nsp E-mail contact: newborn.screening@hhss.ne.gov
E-FAX: (402) 742-2332 Regular Fax: (402) 471-1863

Nebraska Newborn Screening & Genetics Program
Lifespan Health Services, Division of Public Health, DHHS
P.O. Box 95026
Lincoln, NE 68509-5026

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

* *

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

Jeffrey Hoffman, MS, CCC-A, Early Hearing Detection and Intervention (EHDI) Program Manager
(402) 471-6770 Program planning, evaluation and management, systems development
Kerry Julian, EHDI Program Staff Assistant (402) 471-3579
Follow-up, patient education materials distribution, data management, special projects
Mike Rooney, Administrative Assistant (NNSP and EHDI) (402) 471-9731
Meeting planning, patient education materials, translations
Jim Beavers, Business Analyst, EHDI Program (402) 471-1526
Data system planning and testing, development of reports, system security, training and technical assistance.

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

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