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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 2008, newborn screening efforts resulted in successfully identifying and treating 46 newborns affected with conditions in time to prevent problems associated with them:**

- ❖ 1 baby with profound biotinidase deficiency, 3 babies with partial (treated) deficiency
- ❖ 1 baby with congenital adrenal hyperplasia
- ❖ 11 babies with congenital primary hypothyroidism and 1 with compensated hypothyroidism
- ❖ 15 babies with cystic fibrosis
- ❖ 4 babies with hemoglobinopathies (3 sickle cell disease, and 1 Sickle Hgb-C disease)
- ❖ 4 babies with MCAD (Medium Chain Acyl-CoA Dehydrogenase Deficiency)
- ❖ 1 baby with carnitine deficiency due to Maternal Glutaric Acidemia I
- ❖ 1 baby with mild 3-MCC (3-methylcrotonyl carboxylase deficiency)
- ❖ 1 with Methylmalonic Acidemia
- ❖ 1 with Hypertyrosinemia of prematurity
- ❖ 1 with Homocystinuria
- ❖ 1 with mild VLCAD (Very Long Chain Acyl-CoA Dehydrogenase Deficiency)

**The incidence rate of conditions in Nebraska based on the screened conditions identified from 2006 -2008 and number of births screened those three years:**

**1:622 births**

**WHAT IS NEWBORN SCREENING?**

Newborn screening programs have been around for over four decades in all 50 states and in several countries. The compulsory screening panel varies from state to state but the overall goal is the same: prevent or minimize the serious effects of the conditions screened. In 2008 Nebraska’s screening panel required by Neb.Rev. Stat. §§71-519 through 71-524 and administered through regulations 181 NAC 2 included 28 metabolic, endocrine, hematologic and other conditions.

The effects of screened conditions if not detected and treated can range from brain and nerve cell damage resulting in severe mental retardation, to damage to the child’s heart, kidney, liver, spleen, eyes, problems with physical growth, stroke and even death.

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 Cystic Fibrosis (CF) Center is always recommended.

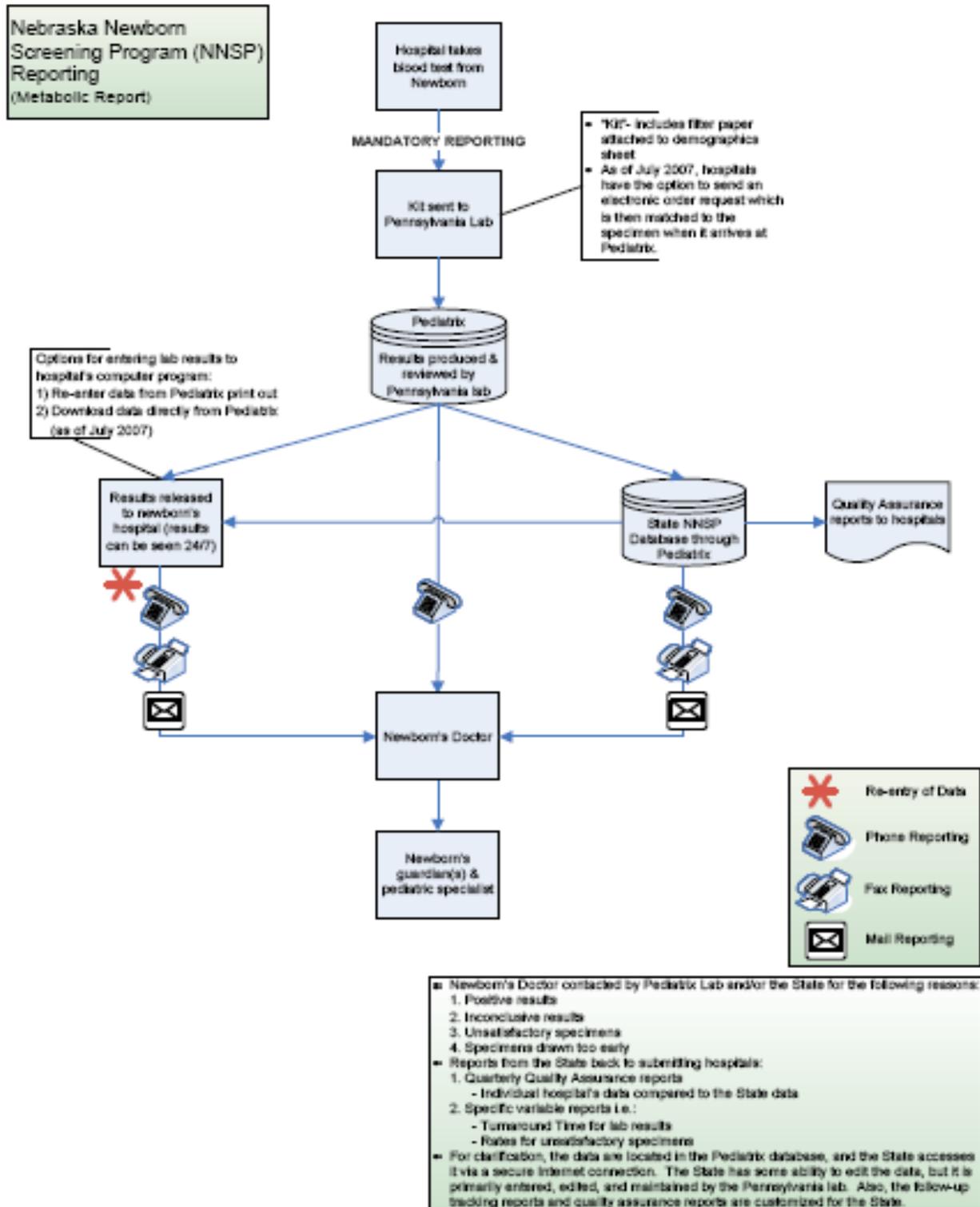
Conditions included in Nebraska’s required blood-spot screening panel in 2008 were:

<b>Argininosuccinic Acidemia</b>	<b>Long Chain Hydroxyacyl-CoA Dehydrogenase Def.</b>
<b>Beta-ketothiolase Deficiency</b>	<b>Medium Chain Acyl-CoA Dehydrogenase Deficiency</b>
<b>Biotinidase Deficiency</b>	<b>Methylmalonic Acidemia (Mutase)</b>
<b>Carnitine Uptake Defect</b>	<b>Methylmalonic Acidemia (Cbl A &amp; B)</b>
<b>Citrullinemia</b>	<b>Multiple Carboxylase Deficiency</b>
<b>Congenital Adrenal Hyperplasia</b>	<b>Phenylketonuria</b>
<b>Congenital Primary Hypothyroidism</b>	<b>Propionic Acidemia</b>
<b>Cystic Fibrosis</b>	<b>Tyrosinemia</b>
<b>Galactosemia</b>	<b>Trifunctional Protein Deficiency</b>
<b>Glutaric Acidemia Type I</b>	<b>Very Long Chain Acyl-CoA Dehydrogenase Deficiency</b>
<b>Hemoglobinopathies</b>	<b>3-Hydroxy 3-Methyl Glutaric Aciduria</b>
<b>(Sickle Cell, Hgb. C &amp; Thalassemias)</b>	<b>3-Methylcrotonyl-CoA Carboxylase Deficiency</b>
<b>Homocystinuria</b>	
<b>Isovaleric Acidemia</b>	
<b>Maple Syrup Urine Disease</b>	

## HOW THE NEWBORN SCREENING PROCESS WORKS

1: TESTING	2: FOLLOW-UP	3: DIAGNOSIS/ INTERVENTION	4: TREATMENT & MANAGEMENT
<p>Baby is born. Dried blood spot specimen is collected @ 24-48 hours of life</p>	<p>Inconclusive or positive screen results reported by phone/fax/letter from lab and follow-up staff to baby's physician.</p>	<p>Depending on the screen result, and on the condition screened:</p> <p>Repeat or confirmatory testing occurs</p>	<p>Once diagnosis is made, treatment begins. (For some life threatening conditions, treatment may occur prior to diagnosis- on recommendation of specialist.</p>
			
<p>Specimen shipped overnight to newborn screening laboratory, Pediatrix</p>	<p>Baby's physician or health care provider contacts baby's parents</p> <p>Parent's bring baby back in</p>	<p>Parent education for signs/symptoms to watch for</p>	<p>Parents receive treatment guidelines / education.</p>
			
<p>Specimen data entered into data system</p>	<p>for evaluation and more testing</p>	<p>Baby's physician consults with and/or refers baby to pediatric sub-specialist appropriate to the condition</p>	<p>Team Support services as appropriate, e.g.:</p>
			<ul style="list-style-type: none"> <li>• metabolic dietitian monitoring &amp; consultation</li> <li>• ongoing blood monitoring</li> <li>• referral to early intervention services</li> <li>• pulmonary/ CF services</li> <li>• pediatric endocrine monitoring</li> <li>• pediatric hematology monitoring</li> <li>• genetic counseling &amp; consideration of family testing</li> <li>• Other allied health services as needed</li> </ul>
<p>Specimen tested for multiple conditions</p> 			

Data flow: This chart demonstrates how data from newborn screening is produced, transmitted and utilized to facilitate the retrieval of newborns at risk for any of the conditions screened, so they can be evaluated, diagnosed and have treatment initiated.



## System Overview

In 2008, 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.



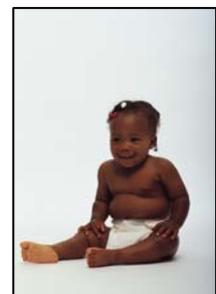
The Newborn Screening Program in the Nebraska Department of Health and Human Services was staffed by Mike Rooney, Administrative Assistant, Krystal Baumert, Follow-up Coordinator, Karen Eveans, Follow-up Specialist, and Julie Miller, Program Manager.

Ongoing consultation with the laboratory, metabolic specialists Richard Lutz, M.D. and William Rizzo M.D., Jill Skrabal, R.D., Kathryn Heldt, R.D., the Cystic Fibrosis Center Director John Colombo, M.D., Dee Aquazzino RN, Coordinator, pediatric endocrinologist Kevin Corley, MD, and pediatric hematologist James Harper, M.D., ensured expert advice and assistance was available as needed throughout the year.

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.



# MAJOR INITIATIVES of 2008 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.
- ❖ The program purchased training materials and distributed a set to every birthing hospital and Children’s of Omaha. This set included Clinical and Laboratory Standards Institute “LA4/A5 Standards for the Collection of Dried Blood Spot Specimens for Newborn Screening,” two laminated posters, and a training DVD, as well as the “Guidelines for Follow-up in Newborn Screening.” This purchase was made thanks to a reduction in cost negotiated between the Association of Public Health Laboratories with CLSI.
- ❖ The Program Manager was one of three presenters in May on an Association of Public Health Laboratories-sponsored webinar on “Public/Private Partnerships” featuring the collaborative relationship between Nebraska’s State-administered Newborn Screening Program with the private laboratory PerkinElmer Genetics Inc.
- ❖ A newborn screening update talk was presented to graduate students in the LEND program at the University of Nebraska Medical Center.
- ❖ Program staff participated in the Heartland Collaboratives’ State-to-State exchange program. Representatives from the Arkansas Newborn Screening Program visited the Nebraska program for a day and a half to learn about follow-up and quality assurance for expanded newborn screening and system management.
- ❖ Internal staff development efforts included the program manager and follow-up staff attending the Association of Public Health Laboratory’s National NBS & Genetics Symposium in San Antonio. Julie Miller presented on the status of the Clinical and Laboratory Standards Institute’s “Guidelines for Newborn Screening for Premature, Sick and Low Birthweight Infants” for which she co-chaired the committee. She also presented a poster on “Policy Analysis of Dried Blood Spot Testing for CMV and Genetic Causes of Hearing Loss,” a joint project of the NBS and Early Hearing Detection and Intervention (EHDI) programs.

## Policy

- Newborn Screening Program staff Krystal Baumert and Karen Eveans served as co-leads on a Heartland Collaborative Project to develop harmonized procedures for follow-up on specimens collected post transfusion. Inter-state transfers of babies between hospitals and NICU’s creates confusion for hospitals, parents and physicians. A goal of this project is to reduce this variability by developing more consistent policies among the eight states in the region.

- The Program Manager continued to serve on regional and national committees: the APHL's Newborn Screening & Genetics Committee providing input on position statements and guidance on dried blood spot storage & use, identification of carriers as a result of newborn screening, quality assurance, contingency planning and other implications from genetic testing. She served on the Heartland Region's Advisory Council, and the Newborn Screening workgroup, and was one of two representatives from the Region on the National Coordinating Centers Long Term Follow-up Work Group.
- The Newborn Screening Advisory Committee (NBSAC) undertook evaluation of proposed revisions to regulations addressing the storage, use and disposal of residual newborn dried blood spots.
- The NBSAC in collaboration with the Advisory Committee for the EHDI program continued its evaluation of the multiple policy implications of various models of integrating dried blood spot testing for Congenital Cytomegalovirus (CMV) and other genetic causes of hearing loss.
- The Newborn Screening Regulations were revised effective July of 2008.
  - previously several of the test results available from the tandem mass spectrometry screen for amino acid, fatty acid and organic acid conditions had been optional. With the Secretary's Advisory Committee's endorsement and Nebraska's Newborn Screening Advisory Committee approval, and these regulation changes, these conditions became part of the mandatory screening test panel.
- The laboratory services contract was awarded to PerkinElmer Genetics Inc., in Bridgeville PA following a competitively bid proposal process and independent review team evaluation of bids. There were two primary changes to the contract. 1) the previously optional/supplemental panel would now be mandatory in accordance with regulation changes, and 2) Saturday pick-up of specimens was to be provided for all hospitals with more than 400 births a year. This expanded the service beyond the Lincoln/Omaha area.

### **Financing Newborn Screening**

The program uses State General funds, the newborn screening fee (\$10/infant) and Title V Maternal and Child Health Block Grant funds to support access to treatment for the metabolic foods and formula. 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 2008 included:

- Blood spot quality issues
- Distribution of Clinical and Laboratory Standards Institute training video on DBS collection
- Lost/missing specimens
- Satisfaction survey/program feedback request

Hospitals were 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 continued to monitor and evaluate increasing rates of unsatisfactory specimens, focusing the "QI hints" educational inserts on this issue.

### **NEWBORN SCREENING ADVISORY COMMITTEE**

A huge debt of gratitude is owed to the dedicated members of the Newborn Screening Advisory Committee who commit their time and expertise to the Nebraska Newborn Screening Program. Much of Nebraska's success can be directly tied to their recommendations and guidance.

The Newborn Screening Advisory Committee (NBSAC) provided technical expertise and policy guidance to the Nebraska Newborn Screening Program. Members commit at least a half a day every three months to advise the State Program. Representatives from PerkinElmer Genetics laboratory regularly provided input, presentations and proposals to the Advisory Committee. Several members provided extensive review and consultation beyond the committee meetings to help the program meet the recommendations of the larger committee.

The members of the NBSAC in 2008 were:

- **Vice-Chair Khalid Awad**, M.D., *Neonatologist*, Neonatal Care PC, Omaha
- **Lawrence Bausch**, M.D., *Neonatologist*, 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
- **CHAIR, James L. Harper**, M.D., *Pediatric Hematologist*, UNMC, Omaha
- **Kathryn Heldt**, RD, *Dietitian*, Children's Hospital Metabolic Clinic, Omaha
- **Mary Kisicki**, RN, *Parent*, Papillion

- **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 are Paid:

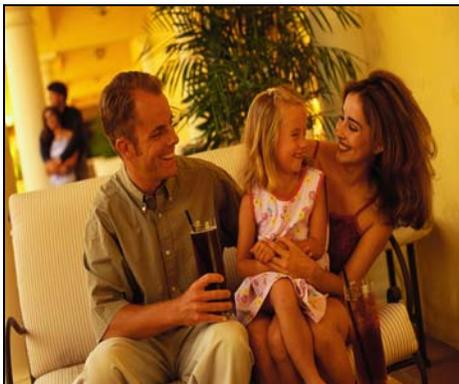


Part of the public health assurance role of newborn screening is ensuring treatment availability and access. Toward that end, the state program manages several contracts to ensure provision of otherwise prohibitively expensive formulas, foods and services not always reimbursed by insurers. Approximately 65 patients received services through these contracts. (Some patients move out of state, new patients move in or are born/and new patients are diagnosed with metabolic conditions).

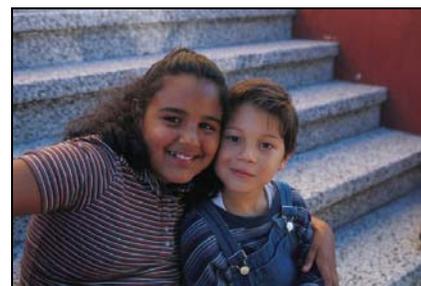
Insurance usually covers medical treatments for some screened conditions, such as prophylactic penicillin for patients with sickle cell disease, or synthetic thyroid hormone for patients with congenital primary hypothyroidism. However, many do not cover the metabolic formulas, and none cover the pharmaceutically 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 the six manufacturers/distributors that have contracts with the state. 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 2008, metabolic formula ordering and distribution and specialized nutritional counseling and monitoring were provided via a contract with the University of Nebraska Medical Center for \$322,827. The individuals eligible for the metabolic foods utilized the pharmaceutically manufactured foods program, ordering foods with a value totaling \$66,948.94.



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. He also works closely with Jill Skrabal, RD to ensure timely contract amendments of appropriate metabolically altered food products as manufacturers continue to expand their offerings.

## 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, and so 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. Therefore, the program

continues to look for sustainable ways to continue to assure access to needed services for people who have these conditions.

### **Nebraska's Newborn Screening Fees**

In 2008 the charge for newborn screening was \$38.50. Of this total charge, the laboratory testing fee was \$28.50 and the state fee (per statute and regulation) was \$10.00 per infant screened. (State fee was used only to help pay for treatment services). Hospital charges are separate. Based on the National NBS & Genetics Resource Center data, of the 47 states that charged a fee for newborn screening in 2008 only 7 were lower (AZ, CT, FL, ID, LA, NC, TX).



## PROCESS/OUTPUT DATA FOR 2008

SPECIMEN  
COLLECTION,  
HANDLING AND  
TRANSPORT



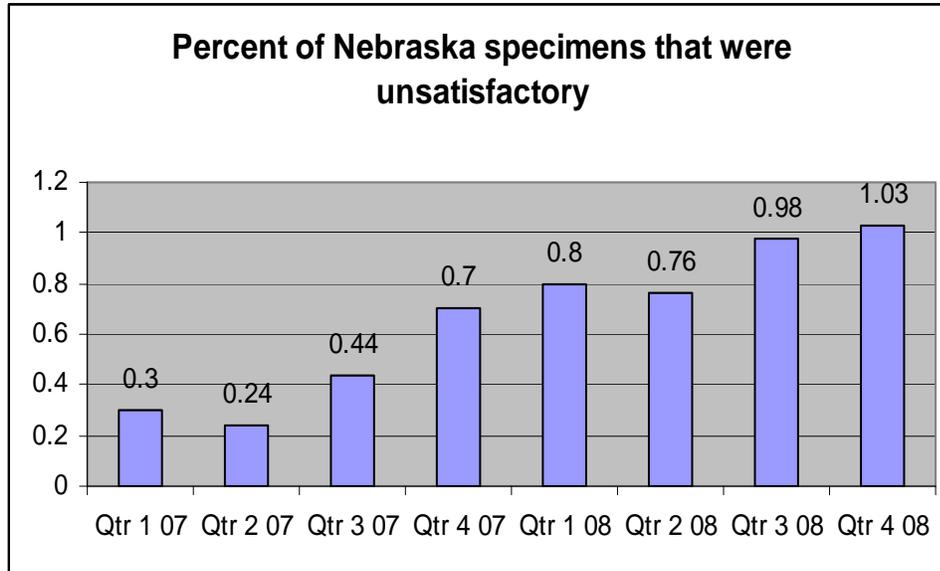
Age at Time of Specimen Collection (Initial Specimen) 2008

Age at time of collection	Number of births	Percent of births
0-12 hours	210	0.77
12-24 hours	130	0.48
Collected day 2 (24-48 hours of age)	25,131	91.92
Day 3	1,551	5.67
Day 4	86	0.31
Day 5	41	0.15
Day 6	21	0.08
Day 7	20	0.07
Over 7 days	149*	0.54
Time of collection unknown	1	0.00

**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.**

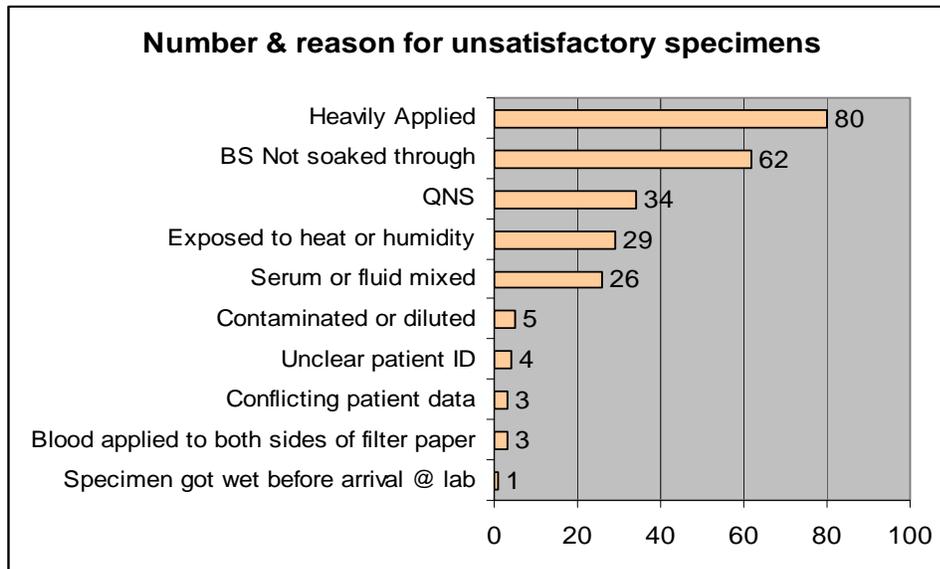
## Unsatisfactory Specimens for 2008

Although Nebraska's unsatisfactory specimen rate was increasing, it was still among the lowest of unsatisfactory rates in the U.S. However, because every unsatisfactory specimen requires the baby to have another specimen collected and creates the potential for a delayed diagnosis, the program takes this issue very seriously. During the last half of 2007 through 2008, despite numerous efforts to address it, the problem with increasing unsatisfactory specimens continued.



### 2008 specific reasons for unsatisfactory specimens

(QNS = quantity not sufficient)



Efforts to address this included:

- Notes to individual hospitals each quarter when their rate significantly exceeds the State average.
- Addressing specimen quality issues in the “Quality Improvement Hints” one-pagers sent with the quarterly individual hospital QA reports (July 2005, Oct 2007, Jan 2008)
- Individual letters to hospital contact persons in the bottom 25<sup>th</sup> percentile of performance (highest 25% of unsatisfactory specimens) sent in January 08, with follow-up progress reports sent during the 3<sup>rd</sup> quarter 08)
- Special letter sent to all Birthing Hospital Risk Managers requesting attention be focused on the unsatisfactory specimens, and identifying liability issues. (Sent in March 2008).
- Training materials (CLSI Standards/Training Video/posters) purchased and sent to every birthing hospital lab director. QI Hints reminder about this sent in April with strong encouragement to use these to address their unsatisfactory rates.
- Pre-summer reminder (each year) about ensuring specimens dry completely to avoid heat denatured specimens.
- Revised Quarterly QA reports to show hospital averages & state averages over time (last 12 quarters) rather than just the most recent quarter.
- In-services were offered at every opportunity with cover letters, and phone conversations regarding specimen quality or turn around times.

**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 2008



## YEAR 2008 SUMMARY OF DRAWN EARLY DATA

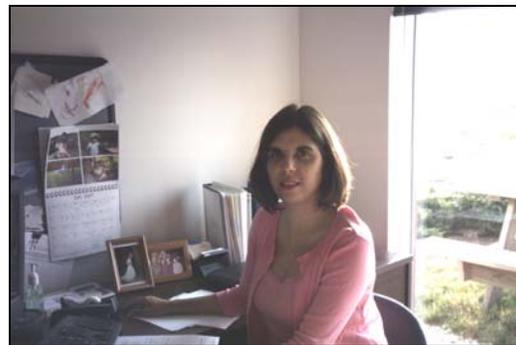
January 1, 2008 – December 31, 2008

#DEs per month:

Jan 2008	21	(18 known transferred)	(1 expired – not repeated)
Feb 2008	28	(25 known transferred)	(1 expired – not repeated)
Mar 2008	29	(23 known transferred)	(2 expired – not repeated)
Apr 2008	32	(24 known transferred)	(3 expired – not repeated)
May 2008	22	(16 known transferred)	(1 expired – not repeated)
Jun 2008	21	(14 known transferred)	
Jul 2008	27	(20 known transferred)	(1 expired – not repeated)
Aug 2008	22	(16 known transferred)	(1 expired – not repeated)
Sep 2008	18	(12 known transferred)	
Oct 2008	29	(11 known transferred)	(1 expired – not repeated)
Nov 2008	20	(9 known transferred)	(1 expired – not repeated)
Dec 2008	26	(17 known transferred)	

**TOTAL:** 295

There were an additional 96 infants that were reported as drawn early and upon notification to the birthing facility it was reported that there was a reporting error made.



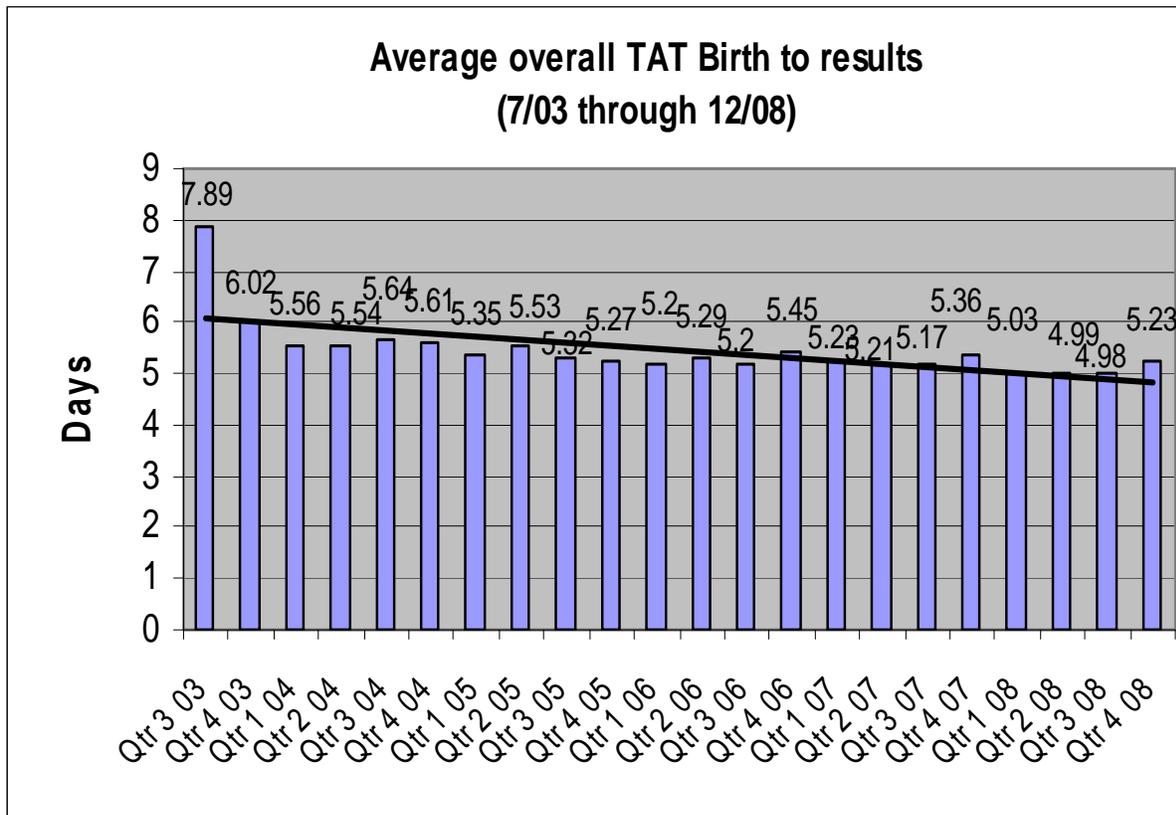
Helping to ensure rapid turn-around times are James DiPerna Manager of MS/MS Laboratory Operations (above left), and Bethany Sgroi M.S.(above right) team leader for the genetic counselors from Perkin Elmer Genetics screening laboratory, who quickly phone the positive or abnormal screening results to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

## Specimen Turn-around 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.

Following the December 2008 holidays, follow-up personnel noticed some urgent positive screening results reported out that had very long turn-around times from birth to the time the result was available. Further investigation revealed systemic problems at a number of facilities with specimens sitting for days at hospital send-out or shipping locations, failure to phone UPS - the shipping company, and other problems associated with non-routine staff persons covering for the usual staff, and a contribution of severe weather across the country impacting shipping times. Notices and reminders were sent to all hospitals.

On the positive side, overall turn around times continued to decline in 2008 thanks to in-lab efforts at PerkinElmer Genetics, and hospital personnel responding to the quarterly quality assurance reports when turn-around times for collection were above the benchmark/average of 1.5 days of age.



## LABORATORY TESTING DATA



PerkinElmer Genetics, Inc., Laboratory uses several instruments to complete the testing. While tandem mass spectrometry provides the screening for 20 of the required conditions, other methods are used for the other eight.

### Presumptive Positive, Inconclusive, & Confirmed Positive Numbers & Rates

#### 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, and 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.

**Most (> 90%) of the babies requiring any follow-up for abnormal results in Nebraska require only a repeat dried blood spot specimen, which usually has a normal result.**

- When a screening result is reported out as “inconclusive,” the recommended follow-up is a repeat dried blood spot specimen. (Most of these will be normal on repeat).
- When a screening result is reported out as “presumptive positive,” the follow-up is treated more urgently and usually a confirmatory test by a different method or on a different kind of specimen (serum, whole blood, urine, etc.) is necessary.

Often the results are abnormal primarily because the baby was premature, sick, low birth-weight, or receiving special treatment such as parenteral nutrition which can interfere with newborn screening results. These babies account for a disproportionate amount of the follow-up needed. However this is not an argument to delay screening on these babies because they are at equal or possibly higher risk of having one of the screened conditions.

- Incomplete data reporting by some State/Territorial programs as of the date of this publication
- National data from the National NBS & Genetics Resource Center notes “Do not use the number of births for rate calculations,” and the number screened is not reported for each condition.

Condition Screened	# Screened	# Presumptive Positive or inconclusive on initial screen	Presumptive Positive Rate	# lost to follow-up	# confirmed Positive/ Diagnosed (classical or partial w tx.)
Biotinidase deficiency	27,021	29	0.10%	0	4
Carnitine Deficiency due to Maternal GA1	27,021	1	0.003%	0	1
Congenital Adrenal Hyperplasia	27,021	38	0.14%	2*	1
Congenital Primary Hypothyroidism	27,021	109	0.40%	1*	12
Cystic Fibrosis	27,021	68	0.25%	6*	15
Galactosemia	27,021	9	0.03%	0	0
Homocystinuria	27,021	93	0.34%	2*	1
MCAD	27,021	6	0.02%	0	4
Methylmalonic Acidemia	27,021	33	0.12%	0	1
PKU	27,021	0	0.00%	0	0
Sickle Cell Disease	27,021	3	0.01%	0	3
Sickle - C Disease	27,021	1	0.003%	0	1
Tyrosinemia (Transient but treated or Hypertyrosinemia of prematurity)	27,021	18	0.06%	0	3
VLCAD	27,021	5	0.01%	0	1
3-MCC	27,021	1	0.003%	0	1
All other abnormal MS/MS results	27,021	139	0.5%	6*	0**

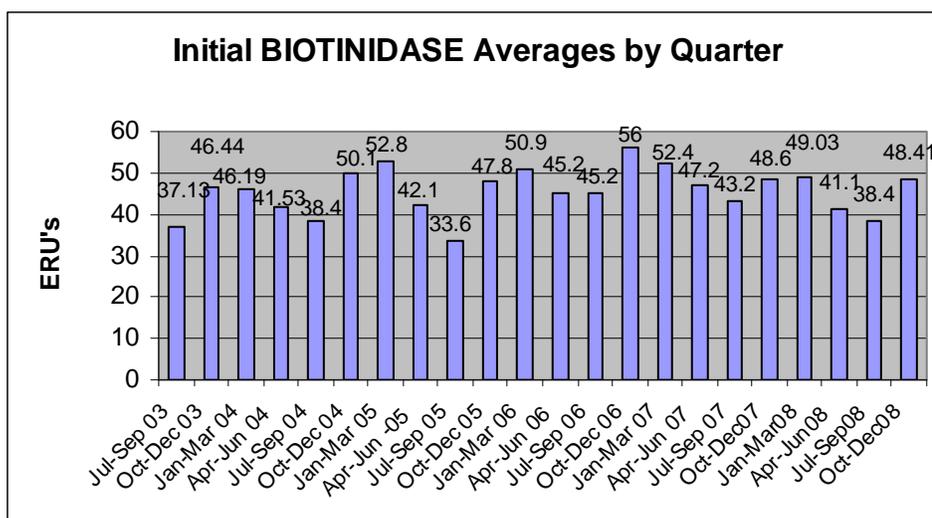
\* all babies lost to follow-up, expired before repeat or confirmatory testing could be completed

\*\* 95 of 139 abnormalities were elevations of multiple amino acids consistent with babies who were receiving parenteral nutrition

## 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 mean averages for biotinidase measures, 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 the amino acids and acylcarnitines used to screen for the fatty acid, amino acid and organic acid disorders are not available from the Tandem Mass Spectrometry results from the screening laboratory. The T4 and TSH results are not included because some results beyond the linearity of the assay would affect the accuracy of this data. These means can reveal something about stability of the assay, reagents etc. over time.

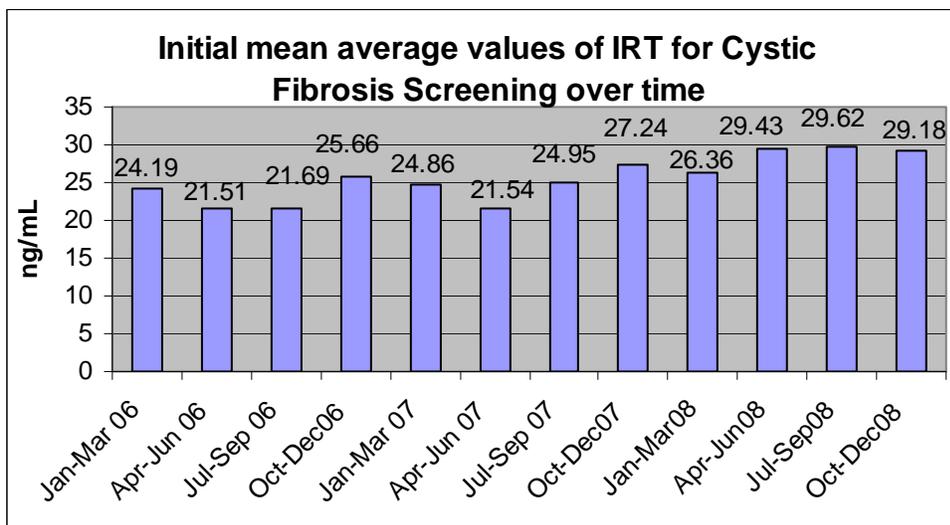
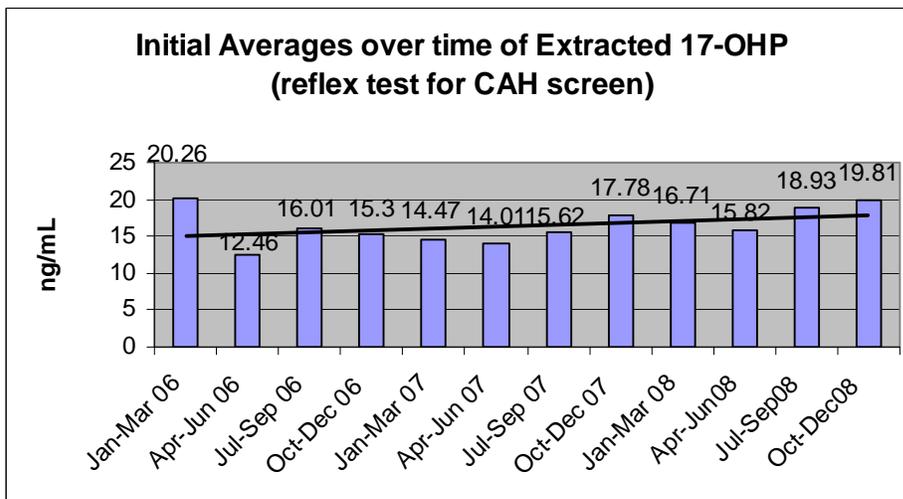
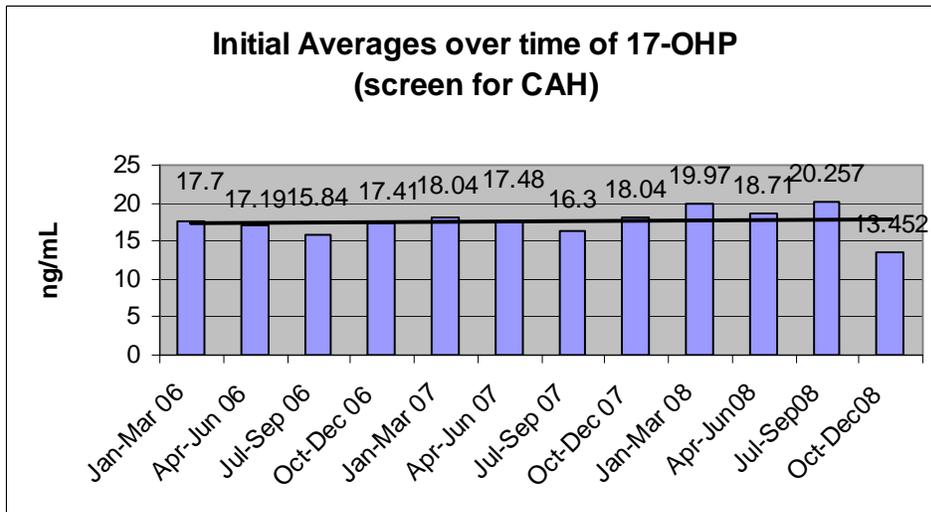
Health care providers familiar with the mean averages might feel more comfortable explaining the “relative risk” to parents of newborns with positive screening results by comparing how far out of range the result is from the mean average and from the normal/expected range.



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 laboratories 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 rejected specimens. In 2008, 29 babies required repeat specimens to re-test enzyme assays used to screen for conditions such as Biotinidase Deficiency and Galactosemia because the initial specimen had been exposed to heat/humidity.

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



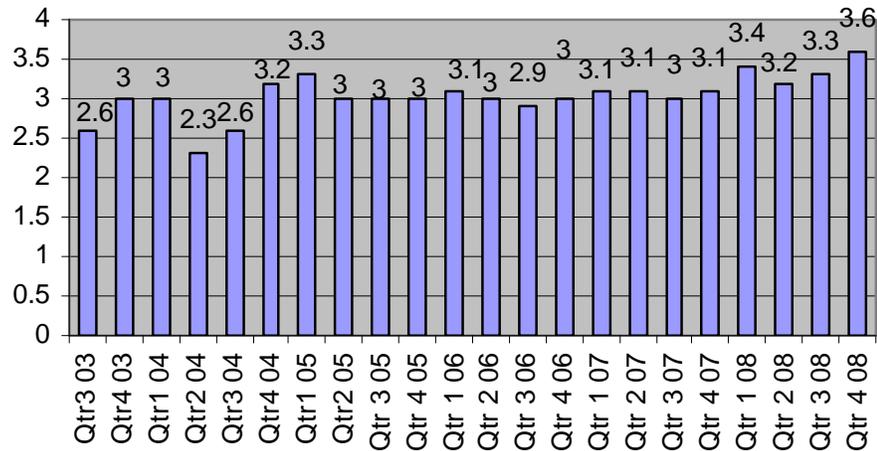
IRTs greater than 90 reflex to test for  $\Delta F508$  the mutation most commonly associated with classical cystic fibrosis. (Those with one copy of the  $\Delta F508$ ) automatically get tested for additional mutations on the INNOGENETICS 36 mutation panel).

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.

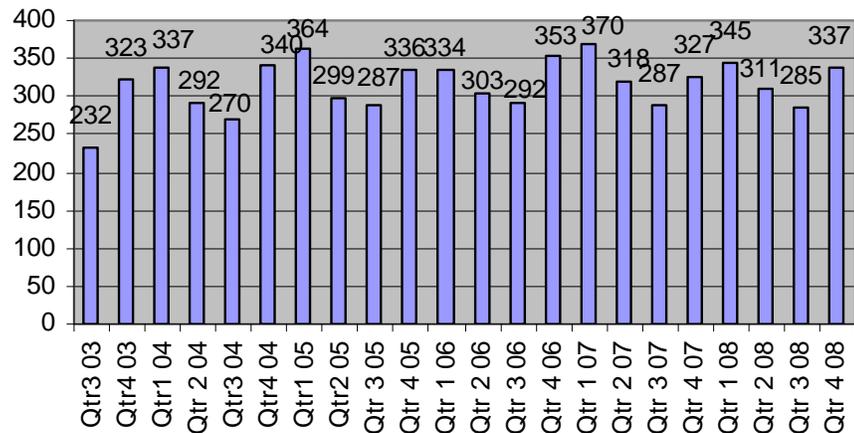
Having this information can mean providing more parents with a bit more peace of mind since most will need only a repeat screen vs. full confirmatory testing.

This also can translate into cost savings because doing a "requested repeat" screen at no charge may be all that is initially recommended vs. more expensive confirmatory testing

**Total Galactose Values mg/dL by quarter**



**GALT Values uM by quarter**



## Out of Hospital / Home Births

In 2008, there were 86 home births reported to the Department of Health and Human Services' Newborn Screening Program (some reported later in 2009). All were screened, except for one of these babies who expired before they could be screened.

## NEWBORN SCREENING DATA

	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008
<b>Total Births</b>	24,209	24,958	25,109	25,515	26,067	26,443	26,349	26,898	27,107	27,094
<b>Births Screened</b>	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	27,021
<b>Total Births Lost to Follow-up</b>	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)		1 (+ 73 not screened as expired @ < 48 hours)
<b>Total Births PP**</b>	357	412	432	456	415	499	503	537	511	553
<b>Home Births</b>	86	109	93	99	70	60	55	69	80	86
<b>Home Births Screened</b>	77	105	88	95	65	60	54	69	78	85
<b>Home Births Lost to follow-up<sup>1</sup></b>	9	4	2 + (3 expired)	2 + (2 expired)	3 + (2 expired)	0	0 + (1 expired)	0	2 (both expired)	1 (expired)

\*Began match with death records beginning in calendar year 2000 to more accurately report #s actually screened.

\*\* PP = Presumptive Positive. Includes all initial screen results requiring either a repeat dried blood spot or another confirmatory specimen and test.

<b>Biotinidase Deficiency</b>	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008
<b>Presumptive Positive</b>	4	2	4	3	4	34*	78	14	5	4
<b>Inconclusive</b>									10	25
<b>Confirmed Negative</b>	2	2	1	1	0	29	71	9	11	23
<b>Confirmed Positive Profound</b>	1	0	0	2	1	0	1	0	0	1
<b>Confirmed Positive (Partial no tx)</b>	0	0	0	0	0	0	0	0	0	0
<b>Confirmed Positive (Partial tx)</b>	1	0	3	0	3	6	5	4	4	3
<b>Lost to follow-up</b>	0	0	0	0	0	0	1**	1**	0	2***

\*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

<b>Congenital Adrenal Hyperplasia</b>	<b>1999</b>	<b>2000</b>	<b>2001</b>	<b>2002</b>	<b>2003</b>	<b>2004</b>	<b>2005</b>	<b>2006</b>	<b>2007</b>	<b>2008</b>
<b>Presumptive Positive</b>	N/A	10	3	17						
<b>Inconclusive</b>									18	22
<b>Confirmed/ Repeated Negative</b>	N/A	9	17	36						
<b>Confirmed Positive</b>	N/A	1	1	1						
<b>Confirmatory or Repeat Lost to follow-up</b>	N/A	0	3*	2*						

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

<b>Congenital Primary Hypothyroidism</b>	<b>1999</b>	<b>2000</b>	<b>2001</b>	<b>2002</b>	<b>2003</b>	<b>2004</b>	<b>2005</b>	<b>2006</b>	<b>2007</b>	<b>2008</b>
<b>Presumptive Positive</b>	108	114	115	129	89	63	58	51	39	57
<b>Inconclusive (drawn early but low T4/high TSH)</b>									20	52
<b>Confirmed Or Repeated Negative</b>	92	104	105	113	75	55	48	41	43	96
<b>Confirmed Positive</b>	13	8	7	15	11	8	9	10	16	12
<b>Confirmatory or Repeat Lost to Follow-up</b>	3	2*	3*	1*	3*	0	1*	0	3*	1

\*Lost to follow-up as babies expired.

Data for Cystic Fibrosis and Hemoglobinopathies are presented in different format because screening for CF is inherently more complex, 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.

<b>Cystic Fibrosis:</b>		<b>2006</b>	<b>2007</b>	<b>2008</b>
<b>Year</b>				
<b>Total Screened Positive</b>		8	3	9
Of those:	Confirmed CF	8	3	9
	Confirmed Atypical CF	0	0	0
<b>Total Screened Inconclusive</b>		62	50	53
Of those:	Confirmed CF	2	3	5
	Confirmed Atypical CF	2	0	0
	Confirmed Carriers	10	12	6
	Found to be within normal limits on repeat	47	32	36
	Expired before confirmation could be done	0	3	6
	Lost to follow-up	0	0	0
	Pending	1	0	0
<b>Total with Meconium Ileus or Bowel Obstruction and positive on DNA</b>		4	14	1
Of those:	Confirmed CF	1	6	1
	Found to be within normal limits	3	7	0
	Pending diagnosis	0	1	0

<b>Galactosemia</b>	<b>1999</b>	<b>2000</b>	<b>2001</b>	<b>2002</b>	<b>2003</b>	<b>2004</b>	<b>2005</b>	<b>2006</b>	<b>2007</b>	<b>2008</b>
<b>Presumptive Positive</b>	13	12	15	5	3	9	1	8	0	0
<b>Inconclusive Repeat rec'd</b>									9	9
<b>Confirmed / Repeated Negative</b>	8	8	9	5	0	6	1	8	8	9
<b>Confirmed Positive (Classical)</b>	0	1	0	0	1	0	0	0	0	0
<b>Confirmed Positive, Duarte (not treated)</b>	3	1 Duarte Hmzgt	0	0	1	0	0	0	0	0

<b>Confirmed Positive, Duarte (treated)</b>	2	2 Duarte Mixed Htrzgt. (1 tx'd 1 year)	6 Duarte Mixed Htrzgt.	0	1	3	0	0	1	
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### Hemoglobinopathy Follow-up Changes:

In 2006 follow-up procedures added the step of sending a reminder letter to the baby's physician before the six-month checkup if the initial confirmatory report indicated a possible alpha, beta or gamma chain variant or combination in the heterozygous state. These typically require additional blood work to diagnose, which previously was not usually reported back to the program. Often these were hemoglobin patterns that had Bart's present on the initial screen and the concern being a possible alpha Thalassemia. 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	FA + Barts: Possible alpha Thalassemia

### Clinically Significant Hemoglobinopathies Confirmed Positive:

	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008
<b>FS</b>	3	2	4	4	5		1	3		3
<b>FC</b>						1	1		1*	
<b>FSC</b>	1	1	2	2		1	1	2	1	1
<b>FE</b>		1						1		
<b>Sickle Beta Thal</b>									1	
<b>Alpha Thal Major</b>			1 (4-gene deletion)							
<b>Beta Thal Major</b>									1	
<b>HPFH</b>							1			
<b>FAE + Beta Thal</b>								6		
<b>FAS + Beta Thal</b>								11		

<b>FAS + Alpha Thal</b>									9		
<b>FAC + Alpha Thal</b>									3		

Dx. = Sickle Hemoglobin C Disease or Hemoglobin C beta thalassemia

**Other Hemoglobinopathies Confirmed Positive in 2008:**

136 Sickle Cell Trait      41 Hgb. C Trait      8 Hgb. D Trait      15 Hgb. E Trait  
 25 Trait + other      3 Alpha Thal silent carrier      6 Alpha Thal Trait      3 Sickle cell trait + alpha Thal trait  
 2 F-Texas      3 miscellaneous trait

Diagnosis unknown for 151 positive Hemoglobinopathy screens (not suspected for clinically significant conditions) (confirmatory results not reported back to program)

<b>MCAD *</b>	<b>1999</b>	<b>2000</b>	<b>2001</b>	<b>2002</b>	<b>2003</b>	<b>2004</b>	<b>2005</b>	<b>2006</b>	<b>2007</b>	<b>2008</b>
<b>Screened Positive</b>	N/A	N/A	N/A	3*	3	5	10	5	0	4
<b>Screened inconclusive (repeat only)**</b>	N/A	2	2							
<b>Confirmed Negative or Repeated normal</b>	N/A	N/A	N/A	2	3	1	7	5	2	2
<b>Confirmed Positive</b>	N/A	N/A	N/A	1	0	4	3	0	0	4

\*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.

<b>Phenylketonuria (PKU)</b>	<b>1999</b>	<b>2000</b>	<b>2001</b>	<b>2002</b>	<b>2003</b>	<b>2004</b>	<b>2005</b>	<b>2006</b>	<b>2007</b>	<b>2008</b>
<b>Presumptive Positive</b>	3	6**	4	3	7**	7	3	6	0	0
<b>Screened Inconclusive (repeat only)***</b>	N/A	N/A	N/A	N/A	N/A	N/A	N/A	N/A	2	0
<b>Confirmed Negative</b>	0	2	2	1	1	1	1	1	2	0
<b>Confirmed Positive Classical PKU</b>	1	1	1	1	2	1	2	0	0	0
<b>Confirmed Positive Hyperphe</b>	2 (tx'd)	1 transient	1	1	3	5 (3 of these tx'd)	0	5 (4 of these treated)	0	0

\*\*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 Screening Results (MS/MS)

MS/MS testing was optional the first six months of 2008 when > 97% of parents opted to receive those results. The full MS/MS panel became part of the required screening panel July 1, 2008.

Including MCAD and PKU (only two required during first half of 2008),  
MS/MS screened conditions on the required screen)

Numbers include a few babies with one abnormality on screen and a different abnormality on repeat.  
Numbers identified as (on repeat) were babies whose first elevated analyte on MS/MS appeared on the second specimen, and all of these were resolved as normal upon further testing.

Initial findings	Out of Range on Screen	Repeated or Confirmed negative	Pending or Lost to Follow-up	Confirmed Positive
Methionine	93 (+26 on repeats)	90	2 expired	1 Homocystinuria
Several Amino Acids	92 (+ 9 on repeats)	87	5 expired	
Several Amino Acids and a generalized elevation of short and medium chain acylcarnitines	3	2	1 expired	
Propionylcarnitine (C3)	13	12		1 Methylmalonic Acidemia
C3 & C3/C2 & C3/C16	20 (+3 on repeats)	20		
Butyrylcarnitine (C4) and other indices such as the relative ratio of C5 to C3	1	1		
Butyrylcarnitine (C4) and other indices such as the relative ratio of C4 to C3	1 (+ 1 on repeat)	1		
Propionylcarnitine (C3) and Butyrylcarnitine (C4)	1 (+1 on repeat)	1		
Butyrylcarnitine (C4)	1	1		
Tetradecenoylcarnitine (C14:1)	3	2		1 Mild VLCAD
Tetradecenoylcarnitine (c14:1) to Palmitoylcarnitine (C16) and other long chain Acylcarnitines	2	2		
3-hydroxydecanoylcarnitine (C10OH), (C8) and (C10)	2 (+ 1 on repeat)	2		
Octanoylcarnitine	6 (+ 2 on repeat)	2		4 MCAD
Glycine and Glutarylarnitine (C5DC)	1	1		
Glutarylarnitine	4 (+1 on repeat)	4		
Tyrosine	18 (+8 on repeats)	15		2 Transient Tyrosinemia (not treated), 1 Hypertyrosinemia of Prematurity
Tyrosine and C3/C2 and C3/C16	1	1		
Methionine an C3, C3/C2, and C3/C16	1	1		
Leucine & Isoleucine	1	1		
Propionylcarnitine (C3) and	1 (+ 1 on repeat)	1		

Butyrylcarnitine (C4)				
Alanine	1	1		
Arginine	(2 on repeats) (tested out of state)			
Isovalerylcarnitine (C5)	1	0	1 expired	
Isovalerylcarnitine & other indices such as the relative ratio of C5 to C4	1	1		
Leucine and Isoleucine	1	1		
Methionine and Tyrosine	5	4	1 expired	
Myristoylcarnitine (C14)	1 (+ 1 on repeat)	1		
3-hydroxyisovalerylcarnitine	1 (+1 on repeat)	0		1 mild 3-MCC deficiency
Free Carnitine & Short Chain Acylcarnitines	1 (+1 on repeat)	0		1 carnitine deficiency due to maternal GA-1
Generalized elevation of short and medium chain acylcarnitines	2 (+ 29 on repeat)	1	1 expired	
<b>2008 TOTALS</b>	<b>279</b>	<b>256</b>	<b>11 expired</b>	<b>12 confirmed positive</b>

\*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 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.**

Several factors can conspire to create delays in treatment, so speed and persistence in follow-up are essential. Some examples of these factors include babies with prolonged treatment in NICU's, parental resistance to confirmatory testing, problems in locating parents because contact information provided to the hospital or recorded on the filter paper collection cards was incorrect or no longer accurate.

Condition & number of babies diagnosed	Average age at intervention/tx.	Range in ages at intervention/tx.
1 Biotinidase Deficiency (profound)	6 days	
3 Biotinidase Deficiency (partial)	17 days	12-25
1 Carnitine Deficiency due to Maternal GA1	27 days	
1 Congenital Adrenal Hyperplasia	22 days (monitored in NICU prior)	
12 Congenital Primary Hypothyroidism	16 days	5-55
1 Compensated Hypothyroidism	33 days	
15 Cystic Fibrosis	18 days	7-42
1 Homocystinuria	20 days	
1 Hypertyrosinemia of Prematurity	29 days	
4 MCAD	5 days	3-8
1 Methylmalonic acidemia	67 days	
1 3-MCC (Mild)	29 days	
3 Sickle Cell Disease	12 days	10-15
1 Sickle Hemoglobin C Disease	14 days	
2 Transient Tyrosinemia	Not treated	

## Nebraska Newborn (Blood-spot) State 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)

# NEBRASKA EARLY HEARING DETECTION AND INTERVENTION ANNUAL REPORT - 2008

## Introduction

Significant hearing loss is one of the most common birth conditions with an estimated incidence 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 soon after birth, the negative impacts can be diminished and even eliminated through early intervention.

In 2000, the Infant Hearing Act established newborn hearing screening in Nebraska. 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.

The Act requires 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.

Newborn hearing screening requires objective physiologic measures to detect hearing loss in newborns and young infants. There are two basic techniques available 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 a diagnostic audiologic evaluation to confirm or rule out a hearing loss and, if hearing loss is present, to identify the type and degree of the loss and to begin intervention services.

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.

## Newborn Hearing Screening Data Reported for 2008

### Birthing Facility Screening Programs

The number of birthing facilities conducting newborn hearing screening increased rapidly from 2000 when only 11 hospitals were conducting either targeted or universal newborn hearing screening (see Table 1). Since 2003, 100% of the birthing facilities in Nebraska have been conducting hearing screenings, consistent with the Neb. Rev. Stat. §71-4742 requirement that a hearing screening test be included as part of the standard of care for newborns. Sixty of the 61 birthing hospitals conduct the hearing screening during the birth admission and one conducts the screening on an outpatient basis following discharge.

**Birthing Facilities Conducting Newborn Hearing Screenings (2000-2008)**

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%
2008	61	61	100%

*Table 1*

### 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 ERS-II data system, an integrated module of the state’s Vital Record system, automatically calculates these figures for each birthing facility.

**Birthing Facility Reports of Required Aggregate Data (2008)**

Number of newborns born in birthing facilities	27,001
Number of newborns and infants recommended for a hearing screening test	25,869
Number of newborns who received a hearing screening during birth admission	26,772*
Number of newborns who passed a hearing screening during birth admission, if administered	25,706*
Number of newborns who did not pass a hearing screening during birth admission, if administered	1066*
Number of newborns recommended for monitoring, intervention, follow-up care	633

\*Includes babies transferred to Children’s Hospital and Medical Center, 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 (25,869 or 95.8%) about newborn hearing screening, hearing loss and normal speech and language development in 2008. 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. There were 82 babies recorded as having been born out-of-hospital in 2008. Parent education material was sent to the parents of the 51 babies who were not admitted to a hospital immediately following birth.

**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 2008 show that 26,772 newborns, or 99.2% of hospital births, were screened during birth admission or prior to discharge to home. The number of newborns screened during birth admission has increased dramatically since reporting began in 2000, when only slightly more than one-third of newborns received a hearing screening during birth admission (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 (2000-2008)**

	2000	2001	2002	2003	2004	2005	2006	2007	2008
Number Receiving a Hearing Screening during Birth Admission	8,978	15,272	22,615	25,275	25,966	26,179	26,615	26,737	26,772
Percent Receiving a Hearing Screening during Birth Admission	36%	61%	89%	97%	98%	99%	99%	99%	99%

*Table 3*

**Newborns Discharged Without a Hearing Screening**

During 2008, the ERS-II reports available for each birthing facility indicated that there were 229 newborns who did not receive a hearing screening during birth admission because the newborn expired prior to screening (103), was discharged to home prior to screening (120) or the parent(s) refused (6).

**Birth Admission “Refer” Rates**

The ERS-II reports available for each birthing facility indicated that 1,066 newborns did not pass the hearing screening during birth admission or prior to discharge to home for those babies who were transferred to another hospital. Of the hearing screenings conducted during birth admission, the “refer” rate for all birthing facilities was 4.0% during 2008 (see Table 4).

**Birth Admission “Refer” Rates (2002-2008)**

	2002	2003	2004	2005	2006	2007	2008
“Refer” rate for birthing facilities	3.7%	3.6%	3.5%	3.4%	3.8%	3.7%	4.0%

*Table 4*

There are two measurement techniques used to conduct newborn hearing screening: Otoacoustic Emissions (OAE) and Auditory Brainstem Response (ABR). Slightly more than 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 two-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 (2008)

	OAE-only	ABR-only	2-Step
Number of Birthing Facilities	32	19*	11
“Refer” Rate	10.3%	1.7%	3.6%

\*includes Children’s Hospital and Medical Center, a non-birthing 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 2008, 633 (59.4% of the babies who did not pass) were recommended for monitoring, intervention and follow-up care by the birthing facilities. Regardless of whether the hospital indicated a recommendation had been made to the parent(s), 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 1,494 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 material was provided by the NE-EHDI Program to the parents of all reported out-of-hospital births during 2008 who were not immediately admitted to a hospital, only 47.6% of out-of-hospital births were screened (see Table 6). The remainder was not screened or the results were not submitted to NE-EHDI Program.

### Out-of-Hospital Births (2001 – 2008)

	2001	2002	2003	2004	2005	2006	2007	2008
Out-of-hospital births	93	99	70	60	55	68	77	82
Number screened	5	16	12	13	15	30	34	39
Percentage screened	5%	16%	17%	22%	27%	44%	44%	48%

*Table 6*

### Confirmatory Testing/Audiologic Data Reported for 2008

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 audiologic evaluation is 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 audiologic diagnostic evaluation. The Advisory Committee’s Audiological Diagnostic Protocol recommends that the outpatient screening facility be prepared to provide comprehensive audiological diagnostic procedures if the outpatient 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-nine (29) of 33 confirmatory testing facilities responded in 2008, representing 83 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	749	143
Number of newborns/infants without a hearing loss	601	45
Number of newborns/infants with a hearing loss	148 (“refer”)	98

*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 the 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 audiologic 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 98 infants were identified with either transient or permanent hearing loss.

### Type and Degree of Hearing Loss

Analysis of the individually-identifiable confirmatory testing reports submitted to the NE-EHDI Program indicates that 42 of the infants with hearing loss meet the criteria for a Permanent Congenital Hearing Loss (PCHL). Twenty seven of the infants with a PCHL were identified

with a bilateral hearing loss, 52% in the mild to moderate range and 48% in the severe to profound range. The remaining 15 infants were identified with a unilateral hearing loss, 67% of which were in the mild to moderate range. Individually-identifiable records indicated that amplification was recommended for 30 of the babies with PCHL and 14 were reported to have been fit with a hearing aid. However, the aggregate reports indicated that 21 infants had been fit with amplification.

**Type and Degree of Permanent Congenital Hearing Loss, 2008 (n = 42)**

Degree ► Type ▼	Bilateral Mild–Moderate	Bilateral Severe–Profound	Unilateral Mild–Moderate	Unilateral Severe–Profound
Sensorineural	11	11	5	5
Conductive	2	-	4	-
Mixed	1	2	1	0
Undetermined	0	0	0	0

*Table 8*

The estimates of the incidence of PCHL in newborns range between 1 to 3 per thousand births nationally. Based on the birth rate in Nebraska during 2008 (27,083 including both birthing facility and out-of-hospital births), an estimated 27 to 81 newborns would be identified with PHL. The incidence of PCHL in Nebraska for babies born in 2008 reported in individual reports is 1.6 per thousand births.

## Tracking and Follow-up Results for 2008

The NE-EHDI Program tracked 1,192 newborns who were reported as not passing a newborn hearing screening during birth admission. Of those 1,066 newborns had a birth admission hearing screening “refer” status and 126 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 birthing facilities for 339 newborns. The aggregate reports from the confirmatory testing facilities indicated that 749 newborns received screenings and 143 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 the NE-EHDI Program, follow-up screening and/or diagnostic evaluations were completed for 1,010 infants with 968 having normal hearing and 42

being diagnosed with a PCHL. The evaluation process is still in progress for 11 infants, the parents of 22 infants refused to complete either the inpatient hearing screening or the recommended follow-up, 14 families moved with no further contact possible, and 13 infants expired before completion of the follow-up services. There were 122 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. These are designed as “Lost to System.”

Diagram 1 tracks the services and outcomes of the 1,192 newborns needing follow-up services through the EHDI system and indicates the results for those infants.

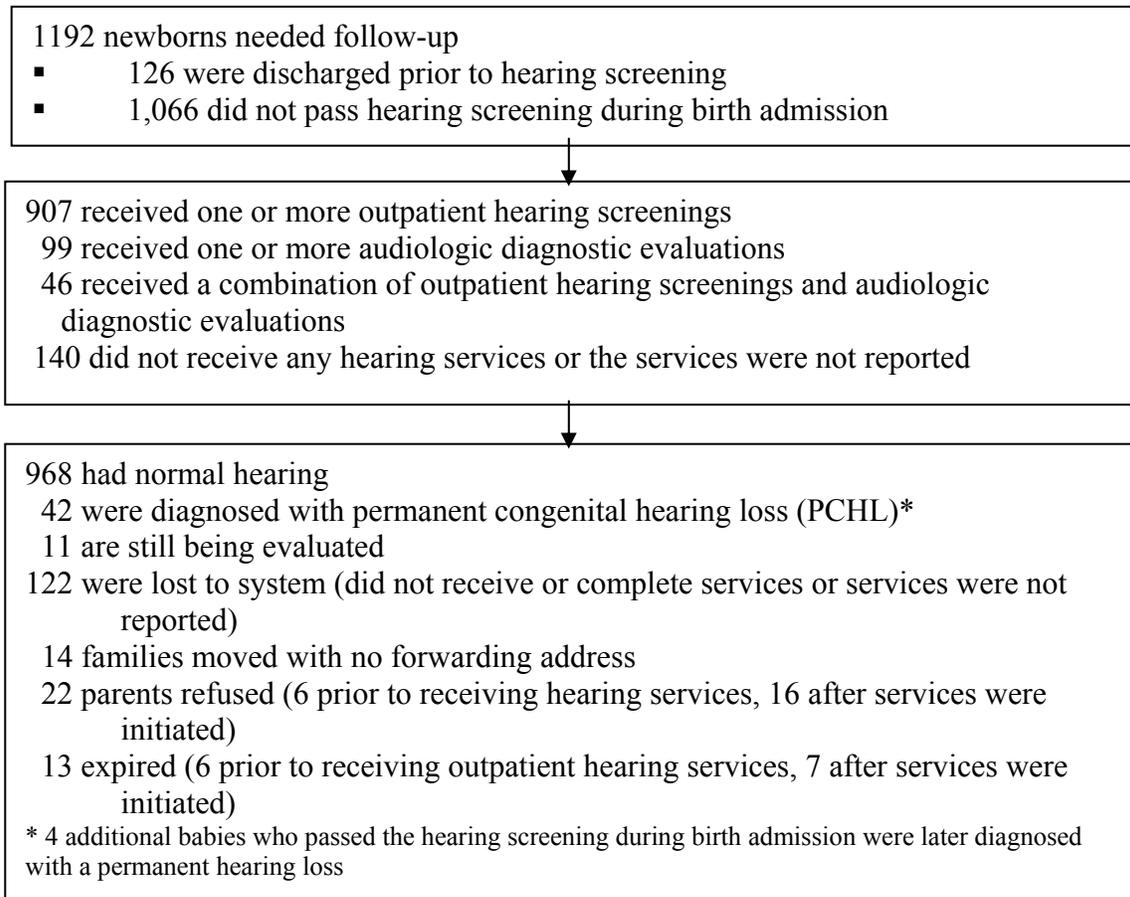


Diagram 1

### Timeliness of Follow-up Screening/Testing

To meet the state and national guidelines of “1-3-6” (hearing screening completed by 1 month, audiologic diagnostic evaluation completed by 3 months, early intervention initiated by six 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, 74.0% received an outpatient screening or diagnostic evaluation prior to one month of age. The peak of follow-up activity occurred at approximately two weeks of age (see Chart 1). The average age of follow-up service initiation was 32.2 days.

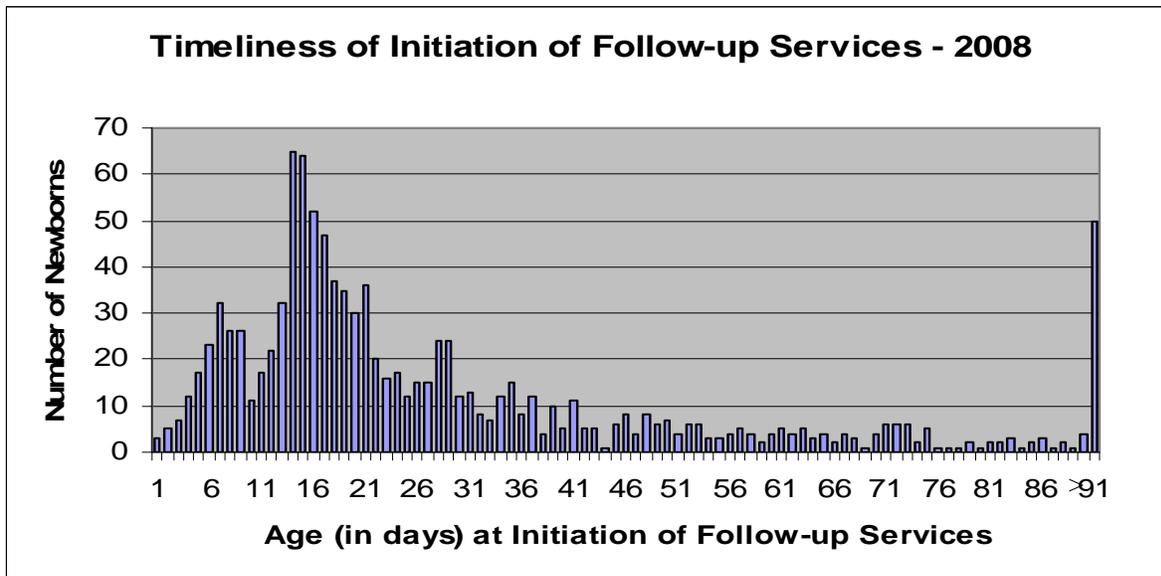


Chart 1

Individual reports were received by the NE-EHDI Program for 46 infants diagnosed with a permanent hearing loss. Four of these were newborns who had passed the newborn hearing screening during birth admission but were diagnosed with a later-onset or progressive hearing loss. The average age at confirmation of hearing loss was 128.5 days with 57% having a hearing loss confirmed before 90 days of age, the recommended benchmark.

### Incomplete Results

Neb. Rev. Stat. §71-4742 states: "...it is the goal of this state to achieve a one-hundred-percent screening rate." While Nebraska continues to make very good progress in developing a comprehensive early hearing detection and intervention system, there are also infants for whom the status of their hearing is not known. Overall in 2008, there were 328 babies whose hearing status has not been objectively established:

- 122 infants with no outpatient follow-up initiated, completed or reported after not passing or receiving the inpatient newborn hearing screening.
- 11 infants 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.
- 14 families moved before hearing services were initiated or completed.
- 6 parents refused the hearing screening during birth admission.
- 16 parents refused the hearing screening after follow-up was initiated but before it was completed.
- 109 newborns expired prior to receiving an inpatient or outpatient hearing screening.
- 7 infants expired before hearing services were completed.

Based on the analysis of the hearing screening and follow-up records, the hearing status (normal hearing or permanent hearing loss) of 98.8% of the 27,083 newborns born in birthing facilities and out-of-hospital has been established.

## Early Intervention

The purpose of the Infant Hearing Act (Neb. Rev. Stat. §71-4735) is to “obtain needed multidisciplinary evaluation, treatment, and intervention services at the earliest opportunity and to prevent or mitigate the developmental delays and academic failures associated with late detection of hearing loss.” Records for the Early Development Network (EDN), Nebraska’s Part C Early Intervention Program, indicate that 38 (90.5%) of the 42 infants born in 2008 with a PCHL were referred to EDN. Of those, 33 verified for special education services. Verification for 27 of the 33 infants was completed prior to six months of age and six infants were verified after six months of age. Four infants were not referred to EDN, six were referred but not verified (three moved or lived out-of-state and the parents of three infants withdrew prior to verification). Thirty-one of the infants diagnosed with a PCHL have an identified medical home.

## ACTIVITIES – 2008

### Funding

The NE-EHDI Program received funding from the Health Resources Services Administration/ Maternal and Child Health Bureau (HRSA/MCHB), the Centers for Disease Control and Prevention (CDC) and the Title V Block Grant. The HRSA/MCHB grant and Title V Block Grant funded the basic operations of the NE-EHDI Program while the CDC cooperative agreement funding supported the development and implementation of the integrated electronic data reporting and tracking system.

### 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 NE-EHDI 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 22 members representing medical, audiology, parents, public health, family support, and education stakeholders. The Advisory Committee met three times during 2008. There are three official sub-committees of the NE-EHDI Advisory Committee: Audiology, Evaluation and Family Support.

## Projects

### **Electronic Data System**

The ERS-II data system was revised to improve functionality and to incorporate additional reports for the birthing facility users. The system was expanded to include the recording of audiologic evaluations and risk factors related to later-onset or progressive hearing loss. The administrative tracking and follow-up system was also further developed.

### **Periodic Early Childhood Hearing Screening**

A Memorandum of Agreement was signed by the Nebraska Head Start Association, the Nebraska Head Start State Collaboration Office and the NE-EHDI Program to support voluntary hearing screening data reporting by Head Start/Early Head Start programs and provision of training and technical assistance by the NE-EHDI Program.

### **Single Point of Entry**

A multidisciplinary workgroup finalized a strategy for a single point of entry to early intervention services for parents of newborns/infants identified with a PCHL. The anticipated outcome of this work is that parents will be able to access timely and appropriate early intervention services through a recognized point of entry, the Early Development Network, with support from staff at the Regional Programs for Students Who Are Deaf/Hard of Hearing who are knowledgeable about hearing loss, its effects on young children, and available resources. Supervisors and services coordinators of the Early Development Network were trained on the single point of entry approach and provided with support materials.

### **Family-to-Family Support**

The Family Support Work Group of the NE-EHDI Advisory Committee provided input for this component of the program. Partnership with the Nebraska chapter of Hands and Voices continued, 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 (NCHALB)**

The NCHALB began providing loaner hearing aids to young children in January 2008. The NCHALB is a partnership between the University of Nebraska - Lincoln Barkley Center, Nebraska Association for the Education of Young Children and the NE-EHDI Program. The NE-EHDI program provides funds to administer the NCHALB and to purchase loaner hearing aids. Requests for additional funding were sent to potential funders. There were 44 hearing aids loaned to 26 young children - 18 with a binaural fitting (two hearing aids) and eight with a monaural fitting (one hearing aid). Funding for permanent amplification was found for eight of the children.

## Summary

- All the current birthing hospitals in Nebraska were conducting newborn hearing screening in 2008. All but one had conducted the hearing screenings during the birth admission.
- The benchmark of 95% of newborns having a hearing screening during birth admission by December 1, 2003, established by Neb. Rev. Stat. §71-4742 continues to be met. In 2008, birthing hospitals reported screening the hearing of 99.2% of newborns during birth admission or prior to discharge to home for those babies who were transferred to another hospital.
- The overall “refer” rate during 2008 for initial hearing screening during birth admission was 4.0%.
- In 2008, follow-up hearing screenings or audiologic evaluations were initiated within one month of birth for 74.0% of those newborns for which follow-up activities were provided.
- The average age at the time of the initiation of follow-up hearing screening or diagnostic evaluation was 32.2 days.
- For the 46 infants identified with a permanent hearing loss, including congenital, later-onset and progressive hearing loss, the average age at confirmation of hearing loss was 128.5 days.
- There are 212 babies (0.8% of those born in 2008) whose hearing was not objectively established and there were 116 who expired before receiving or completing a hearing screening.
- The incidence of Permanent Congenital Hearing Loss identified and reported to NE-EHDI Program (1.6 per thousand in 2008) is within the anticipated range of 1 to 3 per thousand.
- Over 78% of the infants with a permanent congenital hearing loss were verified for early intervention and special education services.

## Nebraska Early Hearing Detection and Intervention Staff



(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)

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