

NEWBORN SCREENING IN NEBRASKA

Newborn Screening for Metabolic and Inherited Disorders

AND

Newborn Hearing Screening



2005 Annual Report

NEBRASKA HEALTH AND HUMAN SERVICES SYSTEM



DEPARTMENT OF SERVICES • DEPARTMENT OF REGULATION AND LICENSURE • DEPARTMENT OF FINANCE AND SUPPORT

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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 disorders 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 2005, newborn screening efforts resulted in successfully identifying and treating 25 newborns affected with disorders in time to prevent problems associated with them:

- ❖ 1 baby with profound biotinidase deficiency, and
 - ❖ 5 with partial biotinidase deficiency
- ❖ 9 babies with congenital primary hypothyroidism
- ❖ 3 babies with hemoglobinopathies (1 sickle cell disease, one sickle hemoglobin- C disease, and 1 HPFH hereditary persistent fetal hemoglobin)
- ❖ 3 babies with MCAD (Medium Chain Acyl Co-A Dehydrogenase Deficiency)
- ❖ 2 babies with classical PKU (Phenylketonuria)
- ❖ 1 baby with Glutaric Aciduria
- ❖ 1 baby with VLCAD (Very Long Chain Acyl Co-A Dehydrogenase Deficiency)

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 disorders screened. In 2005, the gaps between states of what they screened for narrowed with more and more states adopting screening by MS/MS (Tandem Mass Spectrometry). Nebraska has required this MSMS supplementary screening to be offered to every newborn's parents since 2003.

Depending on the disorder, effects can range from brain and nerve cell damage resulting in severe mental retardation, to damage to the child's liver, spleen, eyes, problems with physical growth, stroke and even death.

In the first few days after a baby is born, five drops of blood are collected from a simple heel stick and applied directly to a special filter paper. These blood spots are shipped overnight six days a week to the newborn screening laboratory and tested.

When a specimen is "presumptive positive" for a disorder, the physician is notified and has the infant come back for a repeat or confirmatory test. Once a diagnosis is made, treatment can begin. Treatment varies depending on the disorder and for a few, intervention may be recommended upon learning the initial screening result, prior to obtaining the confirmatory results. Some examples of treatment are: parent education for recognizing signs/symptoms of metabolic crisis, restricting certain foods from the diet, taking a particular vitamin or medication, supplementing a restricted diet with special foods and formula, or preventive antibiotic treatment. Whatever the treatment, the consequences of not beginning treatment in time can be extremely serious for the infant and the family.

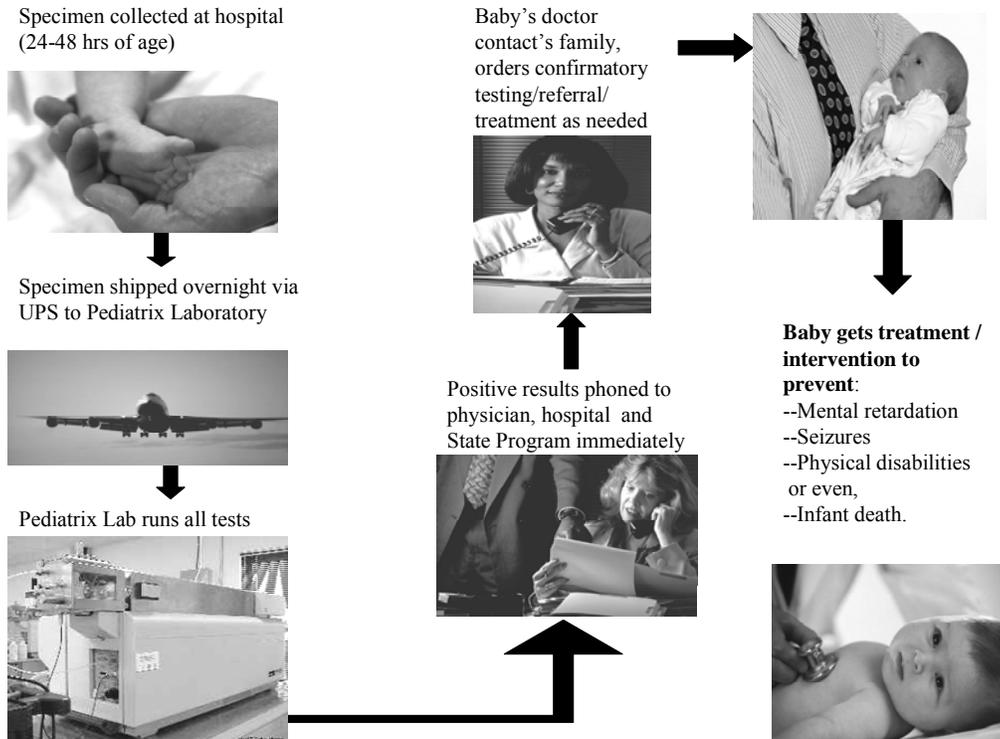
The disorders for which screening is done are individually rare, so consultation with and/or referral to the appropriate pediatric specialist such as a geneticist, metabolic specialist, hematologist or endocrinologist is always recommended.

Individually each disorder is quite rare. However, collectively as many as one in every 800-1000 babies are diagnosed each year in Nebraska with disorders from the current screening panel!

In 2005, the program worked to implement the Newborn Screening Advisory Committee's recommendations to add Cystic Fibrosis and Congenital Adrenal Hyperplasia to the required screening panel. An additional 10 babies per year are expected to be helped as a result of these additions.

Nebraska's Newborn Screening System

Newborn Blood-spot Screening Process



In 2005, 62 Nebraska birthing hospitals sent specimens to Pediatrix Screening Laboratory. This laboratory is under contract with the State of Nebraska to conduct all of the newborn screens.

In 2005, Nebraska required screening for six disorders:

- Biotinidase deficiency,
- Congenital Primary Hypothyroidism,
- Galactosemia,
- Hemoglobinopathies,
- MCAD and
- PKU.

Regulations also required physicians to offer supplemental screening to the parents of all newborns.

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 of physicians and hospital staff using written materials from the Newborn Screening Program helped parents understand their options. Greater than 95% of parents continued to consent to the supplemental screening in 2005.

The Newborn Screening Program in the Nebraska Health and Human Services System was staffed by Mike Rooney, Administrative Assistant, Krystal Baumert, Follow-up Coordinator, Kristen Strasheim, Follow-up Specialist, and Julie Miller Program Manager. Personnel worked closely with metabolic specialists Richard Lutz, MD and William Rizzo, MD from the University of Nebraska Medical Center for ongoing consultation.

A significant amount of planning and development went into revising regulations and procedures to prepare to add Congenital Adrenal Hyperplasia (CAH) and Cystic Fibrosis (CF) to the required screening panel for 2006. The program worked closely with the Pediatrix Screening Laboratory, Dr. Richard Lutz at the Nebraska Medical Center, and the Nebraska CF Center staff (Dr. John Colombo and Dee Acquazzino, B.S.) to develop screening and follow-up algorithms, and parent and physician education materials. Dr. Mark Wilson (Pediatric Pulmonology) and Dr. Deb Perry (Pathology) of Children's Hospital also advised regarding CF screening and sweat testing. In addition, several training opportunities including Grand Rounds on screening for Congenital Adrenal Hyperplasia and Cystic Fibrosis were provided across the State.

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 MD 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.

T.J's Story... What a difference newborn screening would have made...

“April 14th, 2003 my son was born what we thought to be a healthy 8 pound 6 ounce baby, by c-section. Over the course of the next four weeks our lives were about to change forever. My son started vomiting within the next few days. I was told it was reflux, or I needed to burp him in between. At his two week check he was still vomiting and I was still told it was reflux so we went home and another two weeks went by and all he was doing was eating, crying and vomiting so I finally took him in and said something is wrong. They weighed him and he was 6 pounds 15 ounces. They thought the scale was wrong so they took him to another room and checked again. Still the same, we went to another room and checked again. He was still 6 pounds 15 ounces. So they finally agreed with me something was wrong. The doctor took one look at him and knew he was dehydrated. She didn't know what was going on and said he had to be admitted right away and they would do an ultrasound. We got him in and the ultrasound showed nothing so they sent us to a room and did a blood draw. Within 10 minutes there were 10 doctors and nurses standing outside our room and one comes in and said they needed to repeat his labs to make sure there was no error.



They checked it again and within another 10 minutes they had to rush him to the PICU (Pediatric Intensive Care Unit). His sodium level was 103. Normal is 135-145. They had to bring it up slowly otherwise his brain could hemorrhage, or he could start having seizures. We were so scared. We had no idea what was wrong with our son or what the future was to hold for him. Over the next three days they slowly brought his sodium up to near normal 129-130 and sent him to a regular room. He did end up having a seizure in the hospital and was on seizure medication for two years but he has been

seizure free since then. After two long weeks in the hospital and numerous tests and numerous wrong diagnoses, we had an answer. It was Congenital Adrenal Hyperplasia. We were relieved to have an answer and know that with medication he should be OK, but we would still have to wait and see if he had any brain damage from getting so bad. After we had a diagnosis they then told us that they all were surprised he was alive when we brought him in and we will never know how much longer he could have held on if we didn't take him in that day. I thank God for that every day. If this would have been part of the newborn screen test when he was born he would have never gotten that bad and we would have not had to go through the worst six weeks of our lives wondering if our child was going to survive or not.”

The Nebraska Newborn Screening Program would like to thank Corri Stearnes and T.J. for sharing their story, and for Corri's advocacy for newborn screening to help other families who have children with Congenital Adrenal Hyperplasia.

National Attention on Newborn Screening in 2005

Secretary's Advisory Committee

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 and worked to develop a policy and procedure for evaluating candidate disorders for appropriateness for newborn screening as well as other recommendations to the Secretary of Health and Human Services. The SACHDGDNC has three subcommittees established to assist in evaluating newborn screening systems and recommend priorities and strategies for insuring equity and quality amongst screening programs. The subcommittees are: Laboratory, Follow-up and Education. Nebraska's own Amy Brower, PhD serves on the Committee and the Nebraska Newborn Screening Program Manager has the honor of serving on the Follow-up Subcommittee.

Performance Assessment and Evaluation Scheme (PEAS)

A project to provide states with a self-assessment tool for the newborn screening systems progressed in 2005. The PEAS was published in draft form on the National Newborn Screening & Genetics Resource Center's Web site at genes-r-us@uthscsa.edu. The tool provides performance indicators on all elements of newborn screening including education, specimen collection and handling, laboratory testing, and follow-up. Each performance indicator has at least one or two published references. Further guidance to programs was under development in 2005 in the form of additional guidance, or "next steps" to help states meet the performance indicators. Nebraska plans to participate in a pilot of these performance indicators.

ACMG "ACT" Sheets

The American College of Medical Genetics continued to refine the "ACT" sheets. ACT sheets or "action" sheets are one-page guides to help physicians know what next steps to take upon receiving a positive newborn screening result. Some results are specifically indicative of a particular disorder, for example PKU, while others suggest the possibility of more than one. For example, an elevated C3 on the supplemental screen could indicate possible methylmalonic acidemia, or propionic acidemia. In the meantime Nebraska developed, with the advice of pediatric subspecialists on the Advisory Committee, ACT sheets specific to Nebraska's Newborn Screening Program and made these available on the new and improved Web site at www.hhss.ne.gov/nsp. The NBS Advisory Committee will recommend whether to use the ACMG ACT sheets or the Nebraska ACT sheets.

Parent Education Assessment

Under a cooperative agreement with the Maternal and Child Health Bureau, education and communication experts at the University of Louisiana Shreveport completed an assessment of parent and physicians' educational needs around newborn screening. Basic parent education materials were developed, incorporated into a Newborn Screening Toolkit, and made available to all States. Nebraska is evaluating ways to incorporate the toolkit and meet the educational requirements of the State Law.

Follow-up Guidelines by the Clinical Laboratory Standards Institute

Guidelines for short-term follow-up under development for the first time, were sponsored by the former National Committee on Clinical Laboratory Standards (NCCLS) group. Final Committee reviews, responses and approvals were anticipated in 2006.

MAJOR NEBRASKA INITIATIVES of 2005

Education

- ❖ Additional translations of the patient education materials “Parent’s Guide to Your Baby’s Newborn Screening” and the “Supplemental Consent Form” into French and three Sudanese dialects (Dinka, Nuer and Anuak) were initiated. Completion is anticipated for 2006.
- ❖ The program continued to provide supplies of the “Parent’s Guide” and supplemental newborn screening consent forms to all birthing hospitals and upon request to pediatric and family physician clinics and childbirth educators.
- ❖ In March 2005, the program purchased and distributed to every birthing hospital a copy of a 10 minute video for parent education entitled “NEWBORN SCREENING Protecting Your Baby’s Health” developed by the Health Research Education Trust of New Jersey. The video provides an easy to understand general overview of newborn blood-spot and hearing screening. The video was also made available in Spanish to 21 hospitals with higher percentages of Spanish speaking parents. Some hospitals responded that they would incorporate the videos into their prenatal and postpartum teaching.
- ❖ All of the patient education materials provided to physicians at the time they have a patient with a positive screening result were revised. These are intended to be given to parents at the time they receive the positive screening result, so as to provide them with more information and minimize the amount or intensity of anxiety experienced while waiting for confirmatory/diagnostic test results.
- ❖ The Nebraska Newborn Screening Web site was drastically revised to provide much more information in a more relevant way. The Web site is organized for various target audiences: parents, health care professionals, and hospitals or health care institutions. Physicians and hospitals were notified of the availability of this important resource in July. The ACT sheets advising physicians of recommended next steps following positive screens can also be found on this Web site.
- ❖ To help prepare birthing hospital nursery and laboratory staff for the additions of Congenital Adrenal Hyperplasia and Cystic Fibrosis, the state program provided 10 regional in-services at nine locations across the state in these cities: Grand Island, Kearney, Lincoln, Omaha, Norfolk, North Platte and Scottsbluff.

- ❖ Grand Rounds presentations on newborn screening for CAH and CF were provided by doctors Richard Lutz, MD Pediatric Endocrinology/Metabolism/Genetics, and by the Director of Nebraska's only Accredited CF Center, John Columbo, MD Pediatric Pulmonology. These grand rounds were provided in November at Children's Hospital, and broadcast via the Pediatric Grand Rounds tele-medicine education network to several providers.
- ❖ A physician education newsletter was distributed in December to all pediatricians, family physicians, neonatologists and birthing hospitals responsible for newborn screening and pediatric care of children identified with conditions through newborn screening. Articles provided by Dr.'s Lutz and Columbo provided an excellent overview of CAH and CF. They covered anticipated actions physicians may need to take when faced with a positive or inconclusive screen result, and expected outcomes with screening vs. unscreened children.
- ❖ The NCCLS/CLSI training video on newborn screening specimen collection and handling was distributed to three hospitals on loan by the State program. These birthing facilities are to be applauded for requesting this for training new and existing phlebotomists and medical technicians, to improve the quality of specimen collection at their facilities.
- ❖ The Newborn Screening Program informed all birthing hospital laboratories of a live Web-based Centers for Disease Control (CDC) training opportunity on "Quality Dried Blood Spot Filter Paper Blood Collection." The program received positive remarks back from some hospitals that used this opportunity for staff development and training.

Policy

Regulations Revised

Regulation revisions were initiated and completed in 2005. Two changes were related to adding Congenital Adrenal Hyperplasia and Cystic Fibrosis effective in January 2006. Another important change was to revise the regulatory language related to the required timing of specimen collection for births not attended by a physician (usually home births). This change was designed to eliminate any perceived discrepancy between births attended by a physician and those not attended by a physician. All newborns are required to be screened between 24 and 48 hours of birth.

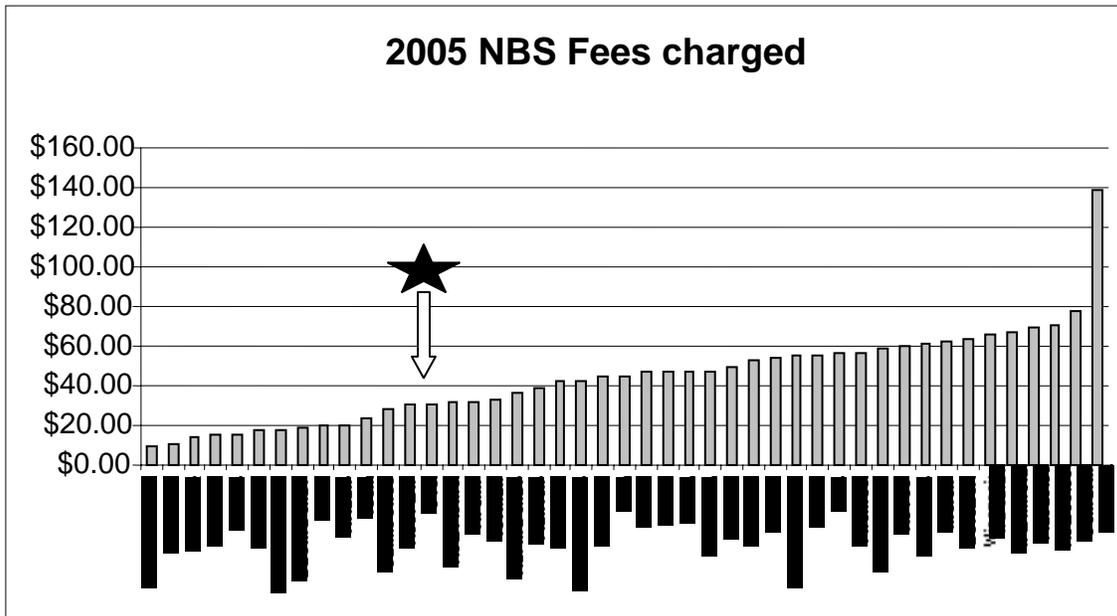
Nebraska Supreme Court Ruling

The Nebraska Supreme Court upheld the constitutionality of Nebraska's Newborn Screening Statute in Douglas County vs. Anaya. In this case the family appealed a Douglas County District Court ruling that their newborn must be screened. In the appeal the Anayas raised a free exercise of religion claim along with a parental substantive due process claim. In the decision the Nebraska Supreme Court unanimously decided that "The State has an interest in the health and welfare of all children born in Nebraska, and the purpose of §71-519 is to protect such health and welfare. This is a rational basis for

the law, and it is constitutional.” The Anayas also argued that the case is moot because the testing was not performed with 48 hours of the baby’s birth. The Court ruled that “one cannot refuse to comply with the testing required within a particular timeframe and then claim that the case is moot because the time has passed.”

Financing Newborn Screening

The Newborn Screening Advisory Committee and Program began focused discussion of alternative means to assure fiscal sustainability. Costs continue to climb while funding streams have been shrinking or staying flat. Nebraska does not have an insurance mandate requiring coverage of the treatments necessary to prevent the mental retardation, physical disability or death for the disorders screened. However, because it is successful, newborn screening continues to identify more people in need of these treatments, which are usually required for the person’s lifetime. The Committee planned to continue to evaluate alternatives for sustainable financing in order to maintain the quality and integrity of the newborn screening system in Nebraska. One alternative considered was increasing the \$10 per infant screened fee that comes back to the State to help pay for treatment. (Laboratory charges were \$20.75 in 2005 in addition to the \$10 fee). According to the National Newborn Screening and Genetic Resource Center data reported by State in 2005, fees for screening ranged from \$0 to \$139.33. Those fees included support for a range of services from laboratory testing only, to laboratory, program administration/follow-up, treatment, education and genetic services. The following tables show state to state comparisons of the 45 states that charged a fee for newborn screening based on the 2005 data:



Source: <http://genes-r-us.us.uthscsa.edu/>

Quality Assurance

Hospital Quality Assurance Reports

Thanks to the outstanding information technology staff at Pediatrix Screening (P.J. Borandi and John O'Rourke), quality assurance reports requested by the State Program were developed to provide hospitals with quarterly feedback on their own performance as compared to statewide averages. The program requested measures of unsatisfactory specimen rates, specimens collected too early (< 24 hours), specimens collected post-transfusion, dissent rates of parents who dissent from the supplemental newborn screen, and average turnaround times from birth to collection, collection to receipt at the lab, in-lab turnaround time and the overall birth to results reporting turnaround time. A different one-page "Newborn Screening Quality Improvement Hints" publication is sent out each quarter with these QA reports.

Ongoing QA Efforts Identified Biotinidase Screening Challenge

Not more than two to three weeks into the summer of 2005, State Newborn Screening Follow-up Coordinator Krystal Baumert recognized a significant increase over usual expected numbers of abnormal newborn screens for biotinidase deficiency. In collaboration with Pediatrix Screening Laboratory, the Program Manager initiated a multi-pronged assessment of the situation to try to determine the cause. The laboratory conducted an intensive evaluation of procedures looking at reagent performance, technician processing, instrument to instrument comparisons, other internal environmental factors and called in the instrument manufacturer to help assess. The Program Manager surveyed other state newborn screening laboratories that utilized the same instrumentation to look for any similar patterns. Concurrently, the program surveyed hospitals to determine specimen handling and shipping procedures to identify any problem areas where specimens may not be consistently dried long enough, or where they were allowed to dry or wait for pick-up in heated conditions. There was no statistical relationship found between those with less than ideal handling and shipping procedures and the numbers of abnormal screens. However the program used this opportunity to follow up with technical assistance to select facilities, and to provide general education to all submitters on proper collection, handling and shipping of specimens. Despite the effort, no single reason could be conclusively determined, although there was some information suggesting that a particular lot of the plates used for processing specimens may have been susceptible to heating specimens, thus resulting in greater numbers of specimens that were heat denatured.

Newborn Screening Advisory Committee

The Newborn Screening Advisory Committee (NBSAC) provides 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. 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 2005, 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 of protocols/algorithms and educational materials:

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 testing protocols, follow-up protocols, follow-up communication tools, physician education, and parent education materials. These tools all needed to be developed to ensure a smooth transition in adding CAH and CF to the required newborn screening panel.

Committee Structure:

The members of the NBSAC in 2005 were/are:

- **Khalid Awad**, MD, *Neonatologist*, Neonatal Care PC, Omaha
- **Lawrence Bausch**, MD, *Neonatologist*, Saint Elizabeth Regional Medical Center, Lincoln
- **John Colombo**, MD, *Pediatric Pulmonologist, Director*, Nebraska Cystic Fibrosis Center, UMC, Omaha
- **Kevin Corley**, MD, *Pediatric Endocrinologist*, Children's Hospital, Munroe/Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Jeanne Egger**, *Parent*, Hallam
- **David Gnarra**, MD, *Pediatric Hematologist*, Children's Hospital, Omaha
- **James L. Harper**, MD, *Pediatric Hematologist*, UNMC, Omaha
- **Kathryn Heldt**, RD, *Dietitian*, Children's Hospital Metabolic Clinic, Omaha
- **Mary Kisicki**, RN, *Parent*, Papillion
- **VICE CHAIR: Richard Lutz**, MD, specialist in *Pediatric Genetics, Endocrinology, Metabolism*, Munroe/Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Bev Morton**, *Parent*, Lincoln
- **Howard Needleman**, MD, *Neonatologist*, Children's Hospital, Omaha
- **Samuel Pirruccello**, MD, *Pathologist*, Regional Pathology Services, UNMC, Omaha
- **Christine Reyes**, MD, *Pathologist*, Pathology Center, Omaha
- **William Rizzo**, MD, 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**, RD, *Dietitian*, Munroe Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Corri Stearnes**, *Parent*, Omaha
- **Douglas Stickle**, PhD, *Technical Director, Clinical Chemistry*, UNMC, Omaha
- **William Swisher**, MD, *Pediatrician*, Lincoln Pediatric Group, Lincoln

- **B.J. Wilson**, MD, *Neonatologist/Perinatologist*, Saint Elizabeth Regional Medical Center, Lincoln, March of Dimes Representative
- **CHAIR Hobart Wiltse**, MD, PhD, *Pediatric Metabolic Specialist*, UNMC, Retired, Omaha,

Dr. Hobart Wiltse served as Chairperson of the Nebraska Newborn Screening Advisory Committee in 2004 and 2005. He has worked tirelessly since the early 1960's in research, education, prevention, treatment and advocacy for quality newborn metabolic screening systems and for patients with metabolic disorders. Thanks in large part to his efforts, Nebraskan's have been rewarded with legislation and systems developed to ensure every newborn benefits from the newborn screen to prevent mental retardation, other physical disabilities and infant death.

Assurance and Cost Effectiveness

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 2005, 58 individuals were served through these programs.

Insurance usually covers medical treatments for some screened disorders 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 disorders 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 \$260,000 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 disorders identified on the supplemental screen that require metabolic formula and foods. Title V Maternal and Child Health Block Grant funds then filled in the gaps for metabolic foods/formula and nutritional counseling.

COST EFFECTIVENESS OF PREVENTION VS. TREATMENT

Data obtained by the NBS program in 2005 identified that generally in Nebraska the prevention costs associated with each patient who has PKU are less than \$10,000 per year.

Remarkably, the treatment costs associated with a patient with severe to profound mental retardation (e.g. a patient with untreated PKU) to receive services in an Intermediate Care Facility for Persons with Mental Retardation such as the Beatrice State Developmental Center are approximately \$101,000 per year, and treatment/services in Community Based Services settings (not including all acute medical care) are greater than \$79,000 per year.

The number of children identified with disorders requiring special formula is anticipated to 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. Therefore the program continues to look for sustainable ways to continue to assure access to needed services for people with the disorders screened.

Makenzie's Story...If only we had known from day one...

“For a new mother there is that unforgettable time in the hospital when you finally get to pull your child away from all the doctors and nurses poking and prodding at your newborn baby and you can finally take a deep breath and enjoy what is now yours. Life couldn't be anymore perfect at that moment. Then it's time to go home!

My wonderful daughter, Makenzie, was born on August 2, 2001. I was a single mother just barely 21 years of age with a lot of plans for the two of us. Only three weeks into life my daughter started becoming very fussy and unsettled. I tried everything to make her happy, I read all the books, called my mother, and of course the doctor. The nurses in the doctor's office assumed that she had colic and only time would heal the aches. By the 4th week my daughter slept only about 10 minutes once every 30 minutes. You can imagine what that would do to a new mom still recovering from childbirth! I was yet unable to get an appointment with her pediatrician so I loaded my crying month old baby up and forced an appointment with the doctor by just showing up. After five minutes of examining Makenzie the doctor said that she must be allergic to her formula. Problem solved, right? At least for a little while...

Shortly after this my daughter gained a little weight and finally started to look like a healthy newborn baby, however, for the next five months we went in and out of the doctor's office, emergency room, and hospital with all kinds of infections. She had chronic sinus infections, bladder infections, kidney infections, RSV, so much I can't even remember.



This life was becoming quite normal to us and every single time, to every single doctor I would ask “What is wrong with my daughter, there is something wrong with her, I know it...what is it”? There were many times when I would show up at a doctor's office or an emergency room pleading for help and finally during one of my hysteric visits at the emergency room, the doctor sent us upstairs to check Makenzie in for the night. That is when we had finally found a doctor that could look at my daughter and say “There is something seriously wrong with this child”.

By this time she had tubes placed in her ears, a muscle biopsy on her leg, an uncounted number of MRI's, CT scans, urinary ultrasounds, and had become visually impaired (cause still unknown), and we had seen more doctors than I ever hope to see again. My little girl had been through more in just six months than most people go through in their entire lives.

We went through the process of being tested for many genetic diseases and finally a doctor had mentioned to me that he believes my daughter has Cystic Fibrosis. I unfortunately had no idea what that meant and to my disbelief the doctor couldn't tell me much more than "your daughter is going to die, the details will be explained to you after we confirm the diagnosis and when we get a CF doctor up to see her". Naturally, I was hysterical and the first chance I had I went to the hospital library. I looked through every book in the place hoping to find a cure for this CF disease but the only thing I kept reading over and over was FATAL.

We waited 10 days in the hospital for the test results and everyday I went through an emotional rollercoaster in disbelief that my baby was going to die...I didn't even have an idea of when. Finally, after the diagnosis was confirmed and the doctors were able to start treatment my daughter was getting better. At this point she had gone completely blind and was very far behind developmentally. She, like many CF patients, had to have a feeding tube placed in her stomach to help with her weight gain. Now she is nearly 5 years old and thriving. Most people have no idea that she isn't a completely normal child.

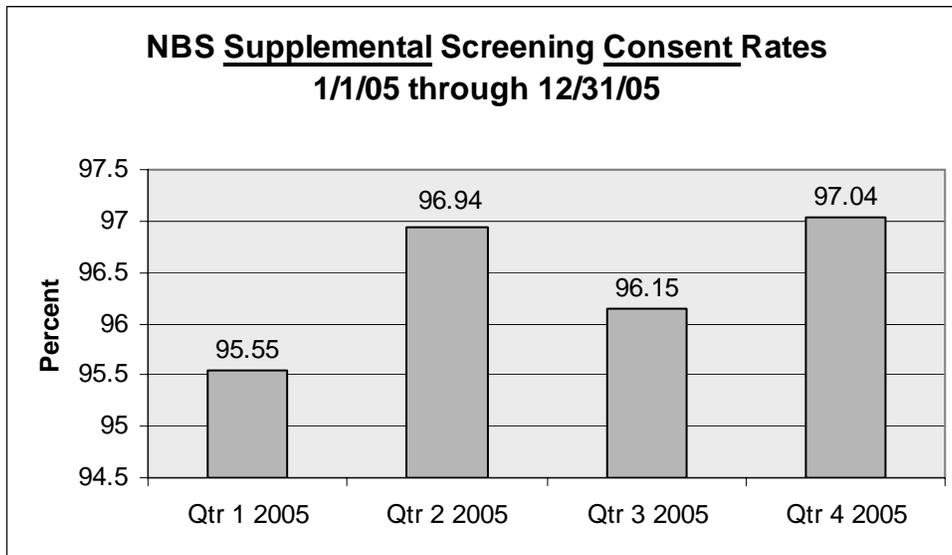
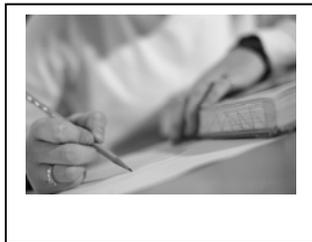
I couldn't tell you how happy I was to hear that CF testing in newborns was beginning in the State of Nebraska. I was even happier to hear that in addition to the testing, there would be information/counseling available to those who test positive. Every time I think about what we went through the first year it takes everything I have to fight away the tears. If only we had known from the beginning, we would have been able to enjoy all those firsts that happen in the first year instead of pleading for an answer."

The Nebraska Newborn Screening Program would like to thank Sara and McKenzie for sharing their story, and for Sara's advocacy to help other families who have children with cystic fibrosis.

PROCESS/OUTPUT DATA FOR 2005

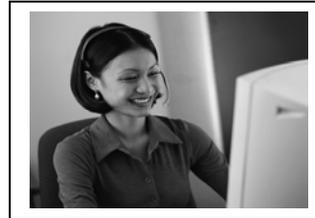
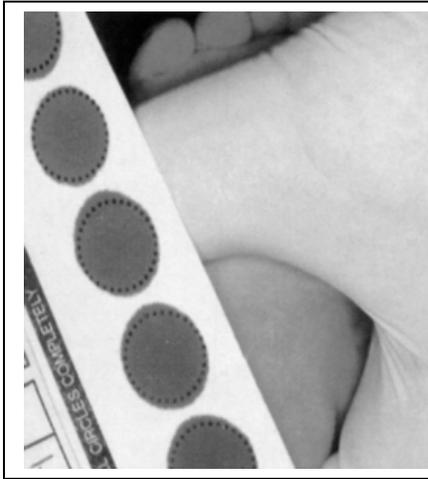
PATIENT EDUCATION

Consent for supplemental screening



Overall for 2005 96.42% of parents consented to the supplemental newborn screening panel from MS/MS. The percent consenting is .86% more than in 2004. Hospital staff report that because it does not require any extra blood, and no additional cost, more parents are requesting it.

**SPECIMEN
COLLECTION,
HANDLING AND
TRANSPORT**



Age at Time of Specimen Collection (Initial Specimen) 2005

Age at time of collection	Number of births	Percent of births
0-12 hours	180	0.68%
12-24 hours	111	0.42%
Collected day 2 (24-48 hours of age)	23,575	88.68%
Day 3	2,247	8.45%
Day 4	232	0.87%
Day 5	47	0.18%
Day 6	22	0.08%
Day 7	21	0.08%
Over 7 days	150	0.56%

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

Unsatisfactory Specimens for 2005

Number of specimens unsatisfactory / Total # initial specimens	159 of 26,288	0.6% of initial specimens
REASONS specimens were UNSATISFACTORY	Number	% of unsats
Exposed to heat or humidity	54	33.9%
Heavily applied, layered or double spotted	45	28.3%
Blood spots not soaked through to the back of the filter paper	23	14.4%
Serum or fluid mixed with sample	13	8.1%
Specimen contaminated or diluted	9	5.6%
QNS (Quantity not sufficient)	7	4.4%
Specimen more than 30 days after the recorded date of collection	3	1.8%
Specimen was wet prior to arrival at lab	3	1.8%

Clotted blood on surface of the filter paper card	1	0.6%
Specimen collection date and time not provided by submitter	1	0.6%
Sample arrived in lab wet	1	0.6%

The art and science of correctly collecting and handling dried blood spots on filter paper requires trained health care professionals with strong skills in attention to detail, 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 2005

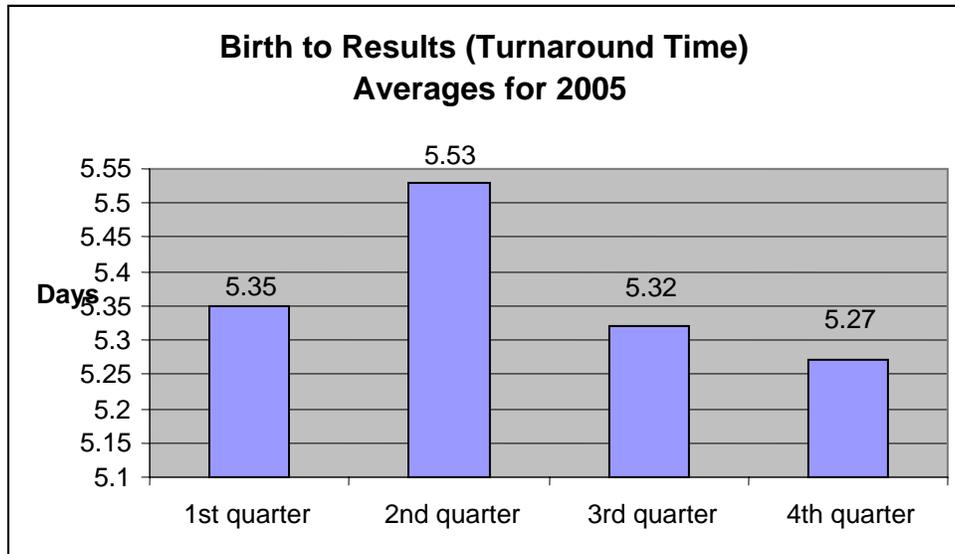


Reason specimen collected at less than 24 hours of age	Number / Percent
Baby to be transferred	85 or 35%
Baby to be transfused	19 or 8%
Unable to determine reason from data received at NNSP	137 or 7%

- ❖ Fourteen of the drawn early newborns did not get repeated as they expired.
- ❖ An additional 117 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.

Specimen Turnaround Time

Regular monitoring of turnaround time of results reporting from the initial specimen, is an important indicator for how well the newborn screening system is functioning to be able to identify affected infants in time to prevent the effects of the disorder.



LABORATORY TESTING DATA

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 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 disorder screened, with the exception of biotinidase screening for which a timely and thorough investigation into the reasons for the unusual numbers of false positives was conducted. (See previous section on quality assurance)

Including only the disorders required to be screened (6), times the number of newborns screened (26,288), the number of tests completed for Nebraska newborns in 2005 were 157,728. Of this 157,728 there were 503 presumptive positive results requiring repeat or confirmatory testing. This is an overall presumptive positive rate of only 0.3%.

Over 96% of Nebraska newborns also received the supplemental Tandem Mass Spectrometry testing for the additional fatty acid, organic acid and amino acid disorders, and only 145 of these required repeat testing (and a handful went on to confirmatory testing).

Specific presumptive positive rates by disorder			
Disorder	National rate 2005*	Nebr. 5 year mean average (2001-2005)**	Nebraska 2005 rates (mean average)***
Biotinidase deficiency	0.15% 15:10,000	0.02% 2:10,000	0.31% 30:10,000
Congenital Primary Hypothyroidism	0.99% 99:10,000	0.35% 35:10,000	0.22% 23:10,000
Galactosemia	0.10% 10:10,000	0.02% 2:10,000	0.01% < 0.5 :10,000
MCAD	0.06% 6:10,000	N/A universal screening began 7/03	0.04% 4:10,000
Phenylketonuria	0.09% 9:10,000	0.01% 1:10,000	0.01% <1:10,000

*National Rate 2005 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. National data source: "2005 National Newborn Screening Report, Initial Screening Results", Biotinidase, Congenital Hypothyroidism, Galactosemia, MCAD, PKU newborns screened total column and newborns presumed with condition column. For Biotinidase Deficiency 23 states reported, CH 31 states reporting, galactosemia 32 states reporting, MCAD 24 states reporting and PKU 31 states reporting as of report run date of 7/5/06. Caution should be used in comparison of numbers.

**Nebraska's 5-year mean: is the mean of the 5 rates figured for each year individually for 2001 through 2005.

***Nebraska's rate 2005: is the number of presumptive positives divided by the total number of newborns screened in 2005.

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 is 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 graphs depict the quarterly mean averages for biotinidase measures, T₄ the primary screen for Congenital Primary Hypothyroidism, 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 Pediatrix Screening laboratory. These means can tell us 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.

	Jan-Mar 2005	Apr-Jun 2005	Jul-Sep 2005	Oct-Dec 2005
Biotinidase mean averages	52.81	42.09	33.55	47.84

	Jan-Mar 2006	Apr-Jun 2005	Jul-Sep 2005	Oct-Dec 2005
T ₄ mean averages	16.226	15.447	16.383	17.075

	Jan-Mar 2006	Apr-Jun 2005	Jul-Sep 2005	Oct-Dec 2005
TSH mean averages	6.765	7.375	6.54	7.068

	Jan-Mar 2005	Apr-Jun 2005	Jul-Sep 2005	Oct-Dec 2005
Galt mean averages	364.9	299.3	287.61	336.94

	Jan-Mar 2005	Apr-Jun 2005	Jul-Sep 2005	Oct-Dec 2005
Total galactose averages	3.3	3	3	3.08

Home Births

For 2005, there were 55 home births reported to the Department of Health and Human Services Regulation and Licensure’s Newborn Screening Program (some reported later in 2006). One of these home births expired and all others (54) were screened. In order to address a Federal District Court ruling, the newborn screening regulations were revised to clearly require all births to be collected within the same time frame (between 24 and 48 hours of birth). Once reported, if the infant is older than 48 hours of age and has not already been screened, the Department now must notify the County Attorney of the need for the infant to be screened.

NEWBORN SCREENING OUTCOME DATA

	1996	1997	1998	1999	2000	2001	2002	2003	2005	2005
Total Births	23,471	23,631	23,862	24,209	24,958	25,109	25,515	26,067	26,443	26,349
Births Screened	23,455 99.9%	23,627 99.9%	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
Total Births Lost to Follow-up	16	4	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)
Total Births PP	356	1,140	547	357	412	432	456	415	499	503
Home Births	78	90	83	86	109	93	99	70	60	55
Home Births Screened	68	86	81	77	105	88	95	65	60	54
Home Births Lost to follow-up¹	10	4	2	9	4	2 + (3 expired)	2 + (2 expired)	3 + (2 expired)	0	0 + (1 expired)

*New match with death records beginning in calendar year 2000, to more accurately report #'s actually screened.

Biotinidase Deficiency	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005
Presumptive Positive	35	5	3	4	2	4	3	4	34*	78*;
Confirmed Negative	34	2	2	2	2	1	1	0	29	71
Confirmed Positive (Profound)	1	1	1	1	0	0	2	1	0	1
Confirmed Positive (Partial no tx)	0	0	0	0	0	0	0	0	0	0
Confirmed Positive (Partial tx)	0	2	0	1	0	3	0	3	6	5

*Screening protocols identified most of these as "inconclusive", for which repeat screening rather than confirmatory testing, ruled out the disorder.

; One lost to follow-up as baby expired.

Congenital Primary Hypothyroidism	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005
Presumptive Positive	276	771	274	108	114	115	129	89	63	58
Confirmed Negative	262	746	265	92	104	105	113	75	55	48
Confirmed Positive	14	10	6	13	8	7	15	11	8	9
Confirmatory Lost to follow-up	0	15	3	3	2*	3*	1*	3*	0	1*

* Lost to follow-up as babies expired.

Galactosemia	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005
Presumptive Positive	9	43	9	13	12	15	5	3	9	1
Confirmed Negative	7	29	9	8	8	9	5	0	6	1
Confirmed Positive (Classical)	0	0	0	0	1	0	0	1	0	0
Confirmed Positive, Duarte (not treated)	1	6	0	3	1 Duarte Hmzgt	0	0	1	0	0
Confirmed Positive, Duarte (treated)	0	6	0	2	2 Duarte Mixed Htrzgt. (1 tx'd 1 year)	6 Duarte Mixed Htrzgt.	0	1	3	0
Confirmed Neg. Classical/CP carrier	1	1	0	0	8	0	0	0	0	0
Confirmatory testing not done¹	0	1	0	0	0	0	0	0	0	0

Hemoglobinopathies		1996	1997	1998	1999	2000	2001	2002	2003	2004	2005
SICKLE CELL DISEASE FS Screened positive		1	3	1	3	2	4	4	5	0	1
Confirmed Positive		1	3	1	3	2	4	4	5	0	1
SICKLE CELL TRAIT FAS Screened positive		16	88	54	120	139	146	156	150	171	186
Confirmed Positive		16	40	54	60	104	102	111 (+1 other variant)	102	81	115
Diagnosis Unknown		N/A	48	0	60	35	44	45	48	90	71
OTHER CLINICALLY SIGNIFICANT Screened positive		-	-	1	3	14	21	2	1	4	6
Confirmed Positive		-	-	1	1	2	3	2	1	2	6
OTHER HEMOGLOBIN VARIANTS Screened positive		-	-	30	228	106	145	150	153	205	162

MCAD *	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005
Screened Positive	NA	N/A	N/A	N/A	N/A	N/A	3*	3	5	10
Confirmed Negative	N/A	N/A	N/A	N/A	N/A	N/A	2	3	1	7
Confirmed Positive	N/A	N/A	N/A	N/A	N/A	N/A	1	0	4	3

*Mandatory screening for MCAD began 7/01/2002. Prior to that about 34% of newborns were voluntarily screened in Nebraska in 2000 and 2001.

Phenylketonuria (PKU)		1996	1997	1998	1999	2000	2001	2002	2003	2004	2005
Presumptive Positive		14	137	43*	3	6**	4	3	7**	7	3
Confirmed Negative		13	106	40	0	2	2	1	1	1	1
Confirmed Positive Classical PKU		1	3	2	1	1	1	1	2	1	2
Confirmed Positive Hyperphe			4		2 (tx'd)	1 transient	1	1	3	5 (3 of these tx'd)	0
Confirmed Positive transient tyrosinemia			24			1	0	0	0	0	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



Tandem Mass Spectrometry Supplemental Screening Results

SUMMARY OF MS/MS FINDINGS Jan 1 – Dec 31, 2005

(Not including MCAD and PKU as these are MS/MS screened disorders 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 Screen	Repeated or Confirmed negative	Confirmed positive	Pending (P) or Lost to Follow-up (LTF)
Tyrosine	29	27		2 LTF
Propionylcarnitine (C3)	9	8		1 LTF
Propionylcarnitine/ Acetylcarnitine/ Palmitoylcarnitine (C3:C2:C16)	12	12		
Propionylcarnitine (C3) & C3:C16 palmitoylcarnitine	1	1		
Generalized elevation of amino acids	42	42		
Ornithine	1			1 LTF
Methionine	18	18		
Methioine & tyrosine	2	2		
C5 OH Hydroxyisovalerylcarnitine	1	1		
Free Carnitine & Short Chain acylcarnitines below normal	3	3		
C5 DC Glutaryl carnitine	1		1- glutaric aciduria	
Butyrlcarnitine C4 & Propionylcarnitine C3	1	1		
Butyrlcarnitine C4	3	3		
Tetradecenoylcarnitine (C14:1)	1		1- VLCAD	
Short & Medium Chain Acylcarnitines	4	4		
Butarylcarnitine C4 & C4:C3 (propionylcarnitine)	1	1		
Dodecanoylcarnitine (C12)	1	1		
C14:1 to C16 ratio and other long chain acycarnitines	1	1		
High concentration of unknown analytes	1	1		
TOTALS	141	134	2	5

*Lost to follow-up designated when the patient/parent can no longer be found, 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 disorder. 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.



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 disorder and shows Nebraska's averages/ranges for 2005. 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 data also includes national averages/ranges according to the most recent available data "National Newborn Screening Report -2005" available at the National Newborn Screening and Genetics Resource Center's Web site:

<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 birth numbers from fewer than 2,000 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 2005 Intervention data	U.S. 2005 Intervention data
Goal age for treatment initiation: Upon Diagnosis	37 States/territories required screening for biotinidase deficiency (+ 1 offered it universally, and 2 offered it to select populations or by request) 35 States reported data.
# diagnosed/treated: 1 profound tx'd @ 18 days 5 partial deficiency treated	33 cases of biotinidase deficiency reported
Mean avg. age at Tx. initiation: 32.66	7 or 21% treated by 7 days of age 4 or 12% treated between 8-14 days of age 4 or 12% treated between 15-21 days of age 10 or 30% treated at > 21 days of age 8 or 24% age of treatment unknown/not reported
Range of ages at Tx. initiation: 18-46 days	Range of ages at Tx initiation.: <3 - > 21 days

Source: <http://www2.uthscsa.edu/nnsis/> data entered as of 7/5/06

Congenital Primary Hypothyroidism

Nebraska 2005 Intervention data	U.S. 2005 Intervention data
Goal age for treatment initiation: As early as possible, upon diagnosis.	52 States/territories required screening for Congenital Hypothyroidism 47 States reported data.
# diagnosed/treated: 9	# diagnosed/treated: 1,879
Mean avg. age at Treatment initiation: 9.22days of age	Age at initiation of treatment: 376 or 20% treated by 7 days of age 622 or 33% treated between 8-14 days of age 241 or 13% treated between 15-21 days of age 342 or 18% treated at > 21 days of age 298 or 16% age at tx. unknown or not reported
Range of ages at treatment initiation: 6-17 days	Range of ages at treatment initiation < 3 to > 21

Source: <http://www2.uthscsa.edu/nnsis/> data entered as of 7/5/06

Galactosemia

Nebraska 2005 Intervention data	U.S. 2005 Intervention data
Goal age for treatment initiation: As early as possible, upon diagnosis. Diet intervention upon positive screening result	52 States/Territories required screening for Galactosemia 46 States reported data
# diagnosed/treated: 0	73 cases of classical galactosemia identified
Mean avg. age at treatment initiation: N/A	Age at treatment: 35 or 48% treated at: 7 days of age or less 17 or 23% treated between: 8-14 days of age 2 or 3% treated between: 15-21 days of age 8 or 11% treated at: >21 days of age 11 or 15% age of treatment: unknown
Range of ages at Tx. initiation: N/A	Range of age at treatment initiation: < 3 days - > 21

Source <http://www2.uthscsa.edu/nnsis/> data entered as of 7/5/06

MCAD - Medium Chain Acyl Co-A Dehydrogenase Deficiency

Nebraska 2005 Intervention data	U.S. 2005 Intervention Data
Goal age for treatment / intervention initiation: As early as possible, upon positive screening result – parent education/consultation.	-34 States/Territories required screening for MCAD -2 States offered testing to select populations or on request -2 States universally offered testing (but not required) (35 reported data)
# diagnosed/treated: 3	# diagnosed/treated: 123
Average age at intervention (avoid fasting): 4.3days	68 or 56% physician notified by 7 days of age 24 or 20% physician notified 8-14 days of age 6 or 5% physician notified 15-21 days of age 4 or 4% physician notified > 21 days of age 19 or 15% age at notification of physician unknown
Average age at treatment (medication): 8 days	34 or 28% treated by 7 days of age 28 or 23% treated between 8-14 days of age 11 or 9% treated between 15-21 days of age 9 or 7% treated at > 21 days of age 41 age of treatment unknown
Range in age at intervention: 4-5 days Range in age at treatment: 5-13 days	Range in age at physician notice: < 3 - > 21 days Range in age at treatment: <3 - > 21 days

Source <http://www2.uthscsa.edu/nnsis/> data entered as of 7/5/06

PKU - Phenylketonuria (Classical PKU)

Nebraska 2005 Intervention data	U.S. 2005 Intervention data
Goal age for treatment initiation: As soon as possible but no later than 7-10 Days after birth.*	52 States/Territories required screening for PKU 45 States reported data
# classical PKU: 2	Cases of classical phenylketonuria 168
Avg. age at treatment: 7.5	51 or 30% treated by 7 days of age 71 or 42% treated between 8-14 days of age 17 or 10% treated between 15-21 days of age 10 or 6% treatment at > 21 days of age 19 or 12% age at treatment unknown or not reported
Range ages at treatment: 7-8 days 50% treated by 7 days of age 100% treated at < 10 days of age	Ranges in ages at treatment: < 3 - > 21 days 30% treated by 7 days of age 47% treated at < 10 days of age

*NIH Consensus Statement October/25/2000: Phenylketonuria: Screening and Management
Source <http://www2.uthscsa.edu/nnsis/> data entered as of 7/5/06

Hemoglobinopathies

Nebraska 2005 Intervention data	U.S. 2005 Intervention data¹
Goal age for treatment initiation: ² 60 days of age or less	51 States/Territories required screening for hemoglobinopathies 1 State offered the testing to select populations or by request 45 States reported data
# cases diagnosed/treated Sickle Cell disease 1 Sickle Hgb. C disease 1	# cases diagnosed/treated Sickle Cell Disease 923 Sickle Hgb. C Disease 498
Mean/Average age (days) at treatment: Sickle Cell Disease 18 Sickle Hgb. C disease 20	195 or 21% Sickle Cell treated 0-30 days 194 or 21% Sickle Cell treated 31-60 days 92 or 10% Sickle Cell treated 71-90 days 67 or 7% Sickle Cell treated > 90 days 375 or 41% Sickle Cell unknown age at of treatment 92 or 18% S Hgb. C Disease treated 0-30 days 99 or 20% S Hgb. C Disease treated 31-60 days 48 or 10% S Hgb. C Disease treated 71-90 days 40 or 8% S Hgb. C Disease treated > 90 days 219 or 44% S Hgb. C Disease unknown age at tx.
Range of ages (days) at treatment: 18-20 100% treated by 60 days of age.	Range of ages (days at treatment) 0 - > 90 Only 41% of cases treated by 60 days of age: (580/1421)

¹ Source <http://www2.uthscsa.edu/nnsis/> data entered as of 7/5/06

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.



PLANS

Screening Panel Expansion: Nebraska now screens nearly 100% of newborns for six disorders and about 95% of newborns for the additional organic acid, fatty acid and amino acid disorders that can be detected on Tandem Mass Spectrometry screening. In 2006 Cystic Fibrosis and Congenital Adrenal Hyperplasia will be screened via the required newborn screening panel. This will make Nebraska's NBS program consistent with the recommendations of the American College of Medical Genetics report of screening for (or universally offering screening for) all disorders in their recommended "uniform" panel and "secondary" panel of disorders.

Other System Planning Efforts: The Nebraska Newborn Screening Advisory Committee will advise the Department of Health and Human Services Regulation and Licensure on implementation of elements of the National Newborn Screening & Genetics Resource Center's "Performance Evaluation and Assessment Scheme." A plan for sustainable funding will be developed, and prioritization to develop three major needs will be done for: disaster preparedness/contingency planning, a comprehensive education plan, and a comprehensive communications plan.

CONTINUING ACTIVITIES

Education: Educational activities from the NNSP will continue through 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. Recommendations for improving the Newborn Screening patient education materials will be sought. The program will work to acquire additional videos for distribution to providers so expecting parents can be exposed to newborn screening during the third trimester of pregnancy. Development of a formal Education Plan will be initiated.

Laboratory Testing: The contract with Pediatrix Screening laboratory is a one year contract, renewable for five years. Annual renewals are dependent on the Department's assessment of contractor's performance. It is anticipated the 4th renewal will occur in 2006. The laboratory will continue to pursue improved efficiencies for screening and collaborate with the State Newborn Screening Program to determine appropriateness for Nebraska of any proposed strategies.

Follow-up, Tracking and Referral: The NNSP will continue to track every newborn to be sure they received an appropriate screen; to follow up on all transferred, drawn early, transfused, unsatisfactory, and presumptive positive specimens; and to facilitate confirmatory testing and referral for diagnostic and treatment services. Review and updating of short-term follow-up procedures will be completed. Until additional resources can be obtained, implementation of any long-term follow-up data collection and analysis designed to inform and improve patient outcomes will be on hold.

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 Newborn Screening Advisory Committee will review the ACMG ACT (Action Sheets) and recommend whether to use these or the Nebraska specific ACT sheets to help physicians in Nebraska know what "next steps" to take when faced with a positive screening result for any of these rare disorders.

Diagnosis: Practitioners are strongly urged to consult with the pediatric specialist appropriate to the disorder for which a newborn has a positive 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 and for which the program with the advice of the NBS Advisory Committee will develop a proposal(s) for sustainable funding. There are some known gaps in treatment/management services particularly for the uninsured or underinsured: e.g. funding for sickle cell and sickle cell trait genetic services, payment sources for routine blood phenylalanine levels for children and women of childbearing age, underinsurance for the number of recommended metabolic clinic visits. 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. Quarterly QA reports and Quality Improvement Hints publications will continue to be sent to individual hospitals for their own evaluation and comparison with statewide numbers.

NEWBORN HEARING SCREENING

Why Is This Report Important?

Significant hearing loss is the most common birth defect with an estimated incidence rate of one to three per thousand live births. Left undetected, hearing loss in infants can negatively impact speech and language acquisition, academic achievement, and social and emotional development. Before newborn hearing screening, many hearing losses were not diagnosed until 2 ½ to 3 years of age. If detected early, however, the negative impacts can be diminished, and even eliminated, through early intervention. Recent studies have consistently shown that children who were identified with a hearing loss later in childhood have delays in the development of speech, language, social and academic skills compared with those identified during the first six months of age.

Newborn hearing screening is an essential preventative 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 requires birthing facilities to educate parents about newborn hearing screening, encourage hospitals to voluntarily begin screening newborns for hearing loss, and by December 2003, include hearing screening as part of the standard of care and establish a mechanism for compliance review. The Act also requires that regulations be promulgated to mandate newborn hearing screening if, by December 2003, less than 95% of newborns in the state were receiving a hearing screening. This report presents the status of newborn hearing screening in Nebraska during 2005 (see Nebraska Newborn Hearing Screening Data for 2005 page 42).

What Is Newborn Hearing Screening?

Newborn hearing screening requires objective physiologic measures to detect hearing loss in newborns and young infants. There are two basic measures that birthing facilities in Nebraska use to screen newborns for hearing loss. Both are easily recorded in newborns and are noninvasive 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 audiological evaluation 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 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.

THE NEWBORN HEARING SCREENING SYSTEM

System Elements

The newborn hearing screening system in Nebraska is composed of five functional elements working together to fulfill the 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, and
- 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 first three principles of the Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs (Joint Committee on Infant Hearing, 2000) are:

1. All infants have access to hearing screening using a physiologic measure. Newborns who receive routine care have access to hearing screening during their hospital birth admission. Newborns in alternative birthing facilities, including

- home births, have access to and are referred for screening before 1 month of age. All newborns or infants who require neonatal intensive care receive hearing screening before discharge from the hospital. These components constitute universal newborn hearing screening (UNHS).
2. All infants who do not pass the birth admission screen and any subsequent re-screening begin appropriate audiologic and medical evaluations to confirm the presence of hearing loss before 3 months of age.
 3. All infants with confirmed permanent hearing loss receive services before 6 months of age in interdisciplinary intervention programs that recognize and build on strengths, informed choice, traditions, and cultural beliefs of the family.

These three major principles serve as the foundation for the screening, referral, and audiological evaluation protocols developed by the Nebraska Newborn Hearing Screening Program (NNHSP) Advisory Committee in 2001. The guidelines established by the NNHSP Advisory Committee are for hearing screening to be completed during birth admission, audiological diagnostic evaluation to begin prior to 6 weeks of age to minimize the need for sedation, and appropriate early intervention activities to be initiated by 6 months of age. The logic model of the NNHSP (see Appendix A) describes the resources and activities needed to produce the projected results of the program.

The five functional elements of the Nebraska Early Hearing Detection and Intervention system are: 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 of 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 Regulation and Licensure through the Nebraska Newborn Hearing Screening 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 staff educate parents to evaluate hearing every six months. *Note:* The Department of Health and Human Services Regulation and Licensure 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. A second inpatient screening is conducted within one to three weeks if the baby "refers" on the first

- screening. The outpatient re-screening for those that “refer” during birth admission may occur at the birthing facility or at a confirmatory testing facility.
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 to the newborn’s Primary Care Provider. Weekly tracking reports are submitted to the NNHSP that identify newborns who “refer,” transfer, or discharge without a hearing screening.
 5. Annual reports are submitted to the NNHSP that indicate the following numbers: 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 second hearing screening should receive an audiological diagnostic evaluation prior to reaching 3 months of age. The purpose of this evaluation is to confirm the presence of a hearing loss and to determine the type and degree of the hearing loss. The primary activities that comprise the confirmatory testing component are:

1. An initial diagnostic evaluation using either OAE or ABR conducted as early as possible after referral, preferably before the infant is 6 weeks old. If the infant “passes” this initial part of the evaluation (outpatient re-screening), no further evaluation is usually needed.
2. If the infant “refers” on the initial part of the evaluation, the testing often proceeds immediately to a comprehensive diagnostic evaluation. This evaluation minimally includes measures of middle ear function (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 NNHSP.
5. Annual reports are submitted to the NNHSP that indicate the number of newborns: who return for follow-up testing, who do not have a hearing loss and 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 pediatric 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. NNHSP notifies PCP about the hearing screening status and need for follow-up evaluation for those that did not pass the inpatient hearing screening or were discharged without a screening.
3. PCP or designee per hospital procedure informs parents of hearing screening results and need for re-screening.
4. PCP (or staff), hospital, or test provider schedules re-screen appointment to be completed in one to three weeks and notifies parents.
5. Provider of outpatient re-screening notifies PCP of results.
6. PCP notifies NNHSP of outpatient hearing re-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, to work with multiple providers to ensure that services are provided, and to become coordinators of services in the future. The recommended protocols for the primary Early Intervention activities within the newborn hearing screening system are:

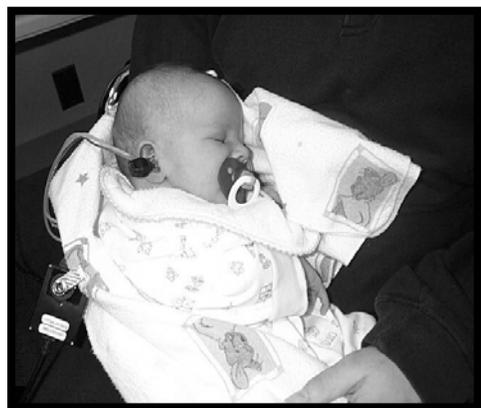
1. PCP or diagnostic evaluator makes referral to Early Development Network (EDN).
2. EDN reviews for eligibility.
3. If eligible, EDN may provide assistance with diagnostic evaluation and treatment.
4. Services Coordinator may facilitate obtaining services from otologists, audiologists, community services, and others.

Tracking and Surveillance

The Nebraska Newborn Hearing Screening Program has been developed based on the requirements identified in the Infant Hearing Act (Neb. Rev. Stat. §71-4735 - §71-4744) and the NNHSP Advisory Committee's recommended protocols 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 NNHSP 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).



NEWBORN HEARING SCREENING DATA FOR 2005

Aggregate Birthing Facilities Data

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 2005, 100% of the birthing facilities in Nebraska were 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 four of the birthing hospitals conducted the hearing screening during the birth admission and one conducted the screening on an outpatient basis following discharge.

Birthing Facilities Conducting Newborn Hearing Screenings (2000-2005)

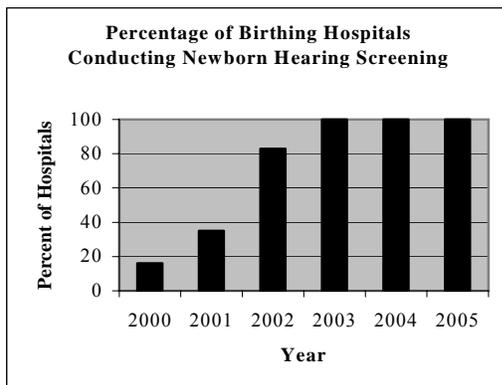


Chart 1

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%

Table 1

Annual Birthing Facilities Reports

Birthing facilities are required to annually report specific information about their newborn hearing screening programs to the Department of Health and Human Services Regulation and Licensure (Neb. Rev. Stat. §71-4739).

Birthing Facility Reports of Required Data (2005)

Number of newborns born	26,252
Number of newborns and infants recommended for a hearing screening test	26,165
Number of newborns who received a hearing screening during birth admission	26,179
Number of newborns who passed a hearing screening during birth admission, if administered	25,256
Number of newborns who did not pass a hearing screening test during birth admission, if administered	908
Number of newborns recommended for monitoring, intervention, and follow-up care	863

Table 2

The data in Table 2 are based on annual aggregate data reported by the birthing facilities. Individual screening results and demographic data are not reported for all births. The NNHSP only receives specific information about newborns that “refer” on the initial hearing screening and about those that were discharged without receiving a hearing screening during the birth admission. The opportunity for error exists within the current manual tracking system due to reporting errors, recording errors, and duplicated entries. Some of these errors were caused by name changes, transfers from birth hospitals to NICUs, and inclusion of newborns who expire in the weekly and/or annual reports. Without a system to accurately determine the status of each newborn’s hearing screening results, errors will be present in spite of the best efforts of everyone involved to provide accurate information.

Parent Education

Recommending a hearing screening test has been operationally defined as educating parents about newborn hearing screening, as required by Neb. Rev. Stat. §71-4740. The NNHSP provides print and video education materials free of charge to hospitals to help fulfill this requirement. Almost all parents (26,165 or 99.7%) received education about newborn hearing screening in 2005.

Newborns Receiving a Hearing Screening

The Infant Hearing Act requires that rules and regulations be adopted and promulgated if at least 95% of the newborns in Nebraska do not have a hearing screening by December 1, 2003, or at any time thereafter. The annual aggregate reports submitted by the hospitals in 2005 show that 99.4% of the 26,349 births registered with Vital Statistics were screened during birth admission. The numbers 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 Chart 2 and 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 (2000-2005)

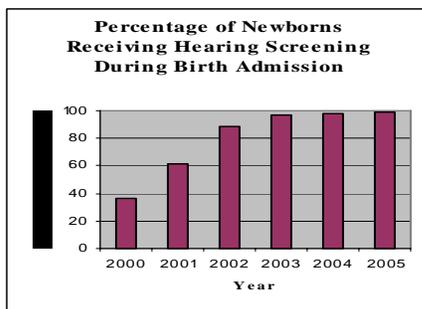


Chart 2

Year	2000	2001	2002	2003	2004	2005
Number	8,978	15,272	22,615	25,275	25,966	26,179
Percentage	36%	61%	89%	97%	98%	99%

Table 3

Newborns Discharged Without a Hearing Screening

During 2005, the annual aggregate hospital reports to NNHSP indicated that there were 255 newborns who did not receive a hearing screening during birth admission because of parent refusal (9), expired prior to screening (101), or discharge prior to screening (145). The NNHSP tracked 974 newborns who were transferred to Neonatal Intensive Care Units in Nebraska and surrounding states prior to receiving a hearing screening. Of those, 12 were later identified with a significant permanent hearing loss.

Birth Admission Refer Rates

The annual aggregate reports received from the birthing facilities indicated that 908 newborns did not pass (“refer”) the hearing screening during birth admission.

Of the newborns with hearing screenings conducted during the birth admission, the refer rate for all birthing facilities was 3.4% during 2005, which compares favorably with national statistics that indicate a refer rate of 3.38% for the 36 states with refer rates of less than 5% during the latter half of 2003 (National Center for Hearing Assessment and Management 2004 State EHDI Survey) and with refer rates for previous years at Nebraska birthing facilities (see Table 4).

Birth Admission Refer Rates (2002-2005)

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

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). 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 approaches, with the OAE-only having the highest refer rate (see Table 5).

Refer Rates for Hearing Screening Techniques (2005)

	OAE-only	ABR-only	2-Step
Number of Birthing Facilities	32	23	10
Number of Newborns Screened	3,709	10,179	12,291
Number of “Refers”	349	324	235
Refer Rate	9.4%	3.2%	1.9%

Table 5

Monitoring, Intervention, and Follow-up

The final aggregate data reported by the birthing facilities is the number of newborns recommended for monitoring, intervention, and follow-up care: 709 (85% of refers) in

2002, 676 (74% of refers) in 2003, 793 (86% of refers) in 2004, and 863 (95% of refers) in 2005.

Aggregate Audiological/Confirmatory Test Provider Data

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

The Advisory Committee for the NNHSP identified the initial level of the follow-up hearing test as an outpatient re-screening of the newborn’s hearing. For those newborns and infants who pass this initial level of the follow-up hearing test, no further audiological evaluation would be needed, unless there are risk factors present that would warrant periodic evaluation. The Advisory Committee recommends that the re-screening occur within the first six weeks to minimize the need to sedate the infant to obtain reliable results and so that intervention can begin early if a hearing loss is identified.

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 referral center should be prepared to provide comprehensive audiological diagnostic procedures if the outpatient re-screening results indicate a “refer” status. However, some communities that do not have pediatric audiology services readily available have opted to have the outpatient re-screening occur at the birthing facility.

Newborn Hearing Screening Annual Confirmatory Testing Facility Reports

Each year data regarding the follow-up hearing tests at confirmatory testing facilities have been gathered by surveying the audiologists in Nebraska. Thirty confirmatory testing facilities responded, representing 63 licensed audiologists. The results of those surveys for 2005 are included in Table 6.

Required Follow-up Hearing Test Data Reported by Audiologists

	Outpatient Re-screenings	Diagnostic Evaluations
Number of newborns/infants receiving a follow-up hearing test	569	141
Number of newborns/infants without a hearing loss	440	56
Number of newborns/infants with a hearing loss	122 (“refer”)	86

Table 6

Rate of Follow-up Outpatient Re-screening and Confirmatory Testing

Since 42 birthing hospitals conducted outpatient re-screenings, those figures are important to present a comprehensive view of the follow-up services being provided in Nebraska. In aggregate reports, the birthing facilities indicated that 384 newborns had received outpatient hearing screenings and confirmatory testing facilities indicated that 569 newborns were re-screened for a total of 953 outpatient re-screenings. With aggregate reporting, it is not possible to determine an unduplicated count, since some infants, especially those with middle ear dysfunction and an accompanying temporary conductive hearing loss, may be screened several times at one or more sites.

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 of 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 NNHSP by audiologists or Primary Care Providers. Statutory authority to require audiologists to report on all newborns and infants that receive audiological evaluations does not exist, so a one-to-one correspondence between the individual results reported to NNHSP and the required annual aggregate reporting does not exist. Audiologists reported conducting 141 diagnostic evaluations of infants born in 2005, identifying 86 infants with hearing loss.

Type and Degree of Hearing Loss

Analysis of the aggregate confirmatory testing reports submitted to NNHSP indicates that 52 of the 86 infants with hearing loss meet the criteria for a Permanent Hearing Loss (PHL). Forty of the infants were identified with a bilateral hearing loss, half of which were in the mild to moderate range and half in the severe to profound range. The remaining 12 infants were identified with a unilateral hearing loss, half of which were in the mild to moderate range and half in the severe to profound range. The aggregate reports indicated that 31 infants had been fit with amplification.

Type and Degree of Permanent Hearing Loss, 2005 (n=52)

Degree ► Type ▼	Bilateral Mild–Moderate	Bilateral Severe–Profound	Unilateral Mild–Moderate	Unilateral Severe–Profound
Sensorineural	18	16	6	6
Conductive	2	-	0	-
Mixed	0	4	0	0

Table 7

The estimates of incidence of permanent hearing loss in newborns range between 1 to 3 per thousand births nationally. Based on the birth rate in Nebraska during 2005 (26,349), an estimated 26 to 80 newborns would be identified with PHL. The incidence of PHL in Nebraska for young children born in 2005 is 1.97 per thousand.

Analysis of Individual Reports Received by NNHSP

The data presented in the two previous sections are based on mandated annual aggregate reports from birthing facilities and confirmatory testing facilities. The individual patient-specific reports received by the NNHSP provide information for additional analysis of the status of the early hearing detection and intervention system in Nebraska. Unduplicated patient-specific information was reported to NNHSP for 858 newborns who referred on birth admission hearing screening, 141 newborns who were discharged to home prior to receiving a hearing screening, 85 newborns/infants who received an audiologic diagnostic evaluation, and 34 newborns/infants who were diagnosed with a PHL. These were the newborns who were tracked through follow-up re-screening, diagnostic evaluations and early intervention.

Follow-up Services

Follow-up screening and/or diagnostic evaluations were initiated for 886 of the 999 newborns who either did not pass the birth admission hearing screening or were discharged without a hearing screening. Of those 886 newborns, follow-up was completed for 839 and was initiated but not completed for 47 of them. There were 113 newborns needing follow-up for whom initiation of follow-up services were not initiated or reported to NNHSP.

In 2005, based on individual reports submitted to NNHSP, there were 109 newborns who needed confirmatory testing beyond the outpatient re-screening to determine the status of their hearing. The results of the 109 who referred on the first outpatient hearing screening were:

- 43 passed on a second re-screening
- 12 were lost to follow-up
- 20 referred on the screening and 1) are in follow-up for conductive hearing loss due to middle ear dysfunction, 2) have not received additional follow-up services, or 3) the results have not been reported to NNHSP
- 34 referred on the first or second outpatient re-screening and then received a diagnostic evaluation

Fifty one (51) newborns received a diagnostic evaluation as the initial step for outpatient follow-up. In addition to those, additional diagnostic evaluations included 34 for infants who did not pass the outpatient re-screenings (see previous paragraph), two for infants who had passed the inpatient hearing screening and one infant born out-of-state but identified while a resident of Nebraska. The results of the 88 diagnostic evaluations are:

- 39 had normal hearing established either initially or following medical management for middle ear dysfunction
- 14 are in follow-up for conductive hearing loss due to middle ear dysfunction or to complete the evaluation
- 1 was lost to follow-up
- 34 were diagnosed with a PHL, including two infants who had passed the newborn hearing screening and one infant born outside of Nebraska. One moved

out-of-state and was categorized as Lost to follow-up. Sixteen of those infants were identified prior to 3 months of age and the average age at identification was 122.4 days.

Diagram 1 tracks the progress of the 999 newborns needing follow-up services through the newborn hearing screening system to the point of hearing status established (normal hearing, permanent hearing loss), results pending, or lost to follow-up.

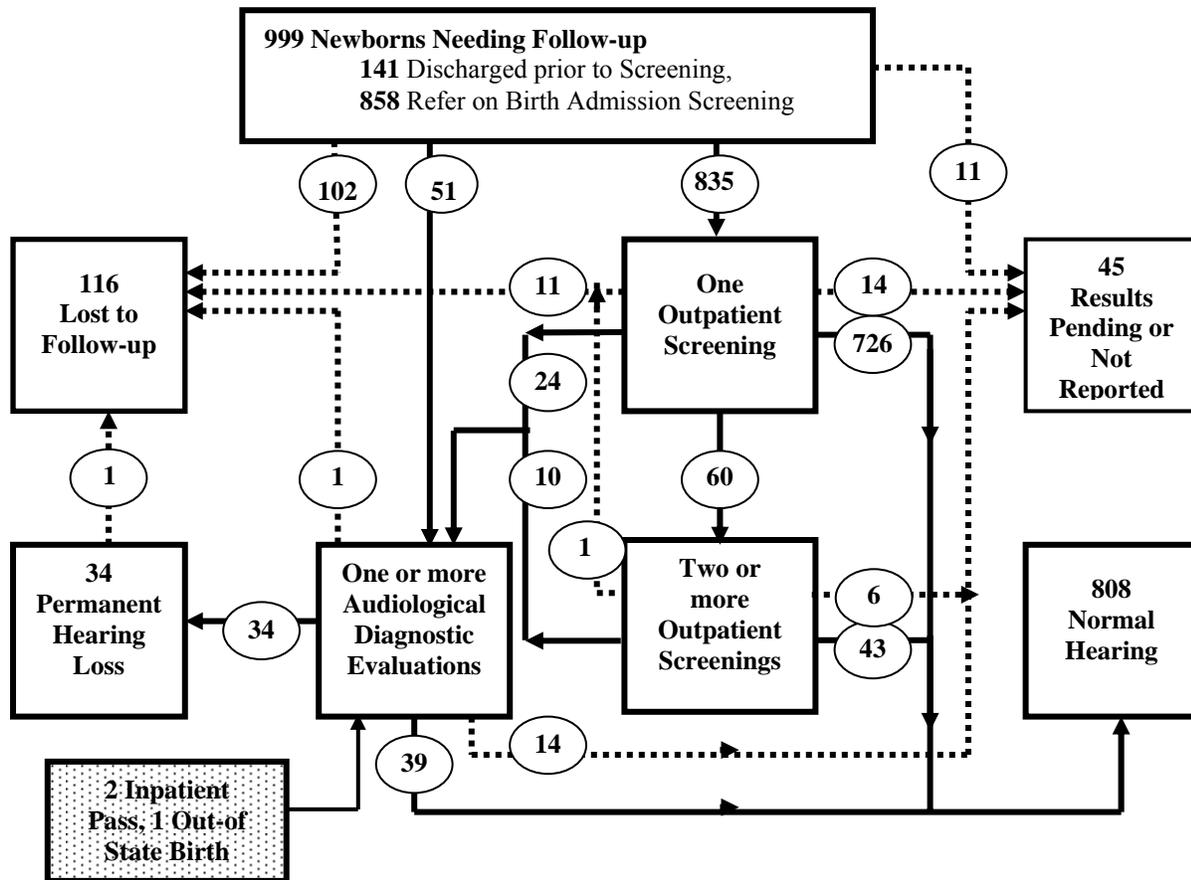


Diagram 1

Timeliness of Follow-up Re-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 of follow-up activities is an important aspect of the quality of services. For the newborns who did not pass the initial hearing screening during birth admission and who received follow-up services, 75.2% received an initial outpatient re-screening or diagnostic evaluation prior to 1 month of age. The peaks of follow-up activity occurred at approximately 1 week, 2 weeks, and 3 weeks of age (see Chart 3). The average age of follow-up service initiation was 29.8 days.

Of the 120 newborns who were discharged prior to screening and received follow-up

services, the average time to the initial outpatient hearing screening was 24.2 days with 103 (85.8%) of those newborns receiving the initial screening prior to 1 month of age. Although an initial outpatient newborn hearing screening by 1 month of age does not meet the intent of Neb. Rev. Stat. §71-4739 for each newborn to be screened during birth admission, it does meet the national Early Hearing Detection and Intervention goal of every newborn having a hearing screening by 1 month of age.

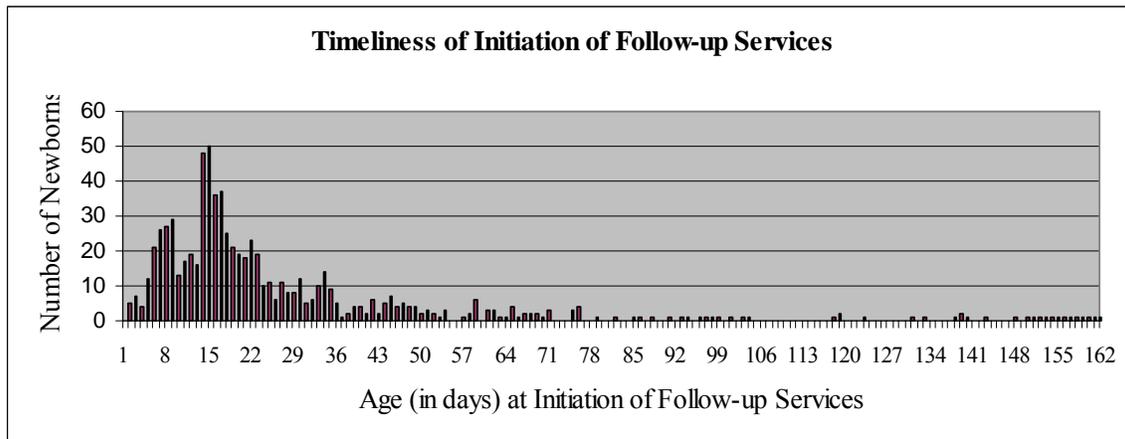


Chart 3

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 newborn hearing screening system, there are also infants for whom the status of their hearing is not known. In 2005, there were at least 311 newborns whose hearing status has not been established:

- 35 were identified with hearing problems but either additional follow-up evaluations have not been completed or the results were not submitted to NNHSP. Of those, 19 had indications of middle ear dysfunction and 14 had received a diagnostic audiological evaluation.
- 3 of the 141 newborns who did not receive a hearing screening during birth admission also did not receive an initial hearing screening as an outpatient or the results were not submitted to NNHSP
- 7 of the 858 who "referred" on the hearing screening during birth admission either had no outpatient re-screening completed or the results were not submitted to NNHSP
- 116 infants were categorized as lost to follow-up (the family moved with no forwarding information, the primary care physician for follow-up communication could not be identified, or repeated efforts to obtain the hearing screening results were not successful)
- 40 of the out-of-hospital births had not been screened or the results were not submitted to NNHSP
- 101 newborns expired prior to receiving a hearing screening

Based on the analysis of the aggregate hospital reports and actual file counts, the hearing status of only 1.2% of the 26,349 newborns was not confirmed as either normal or with a permanent hearing loss present.

Out-of-Hospital Births

Neb. Rev. Stat. §71-4740 requires the Department of Health and Human Services Regulation and Licensure 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. Although parent education was provided to the parents of all reported out-of-hospital births during 2005, only 27.3% (15) of the 55 out-of-hospital births were screened (see Table 8).

Out-of-Hospital Births (2001 – 2005)

	2001	2002	2003	2004	2005
Out-of-hospital births	93	99	70	60	55
Number screened	5	16	12	13	15
Percentage screened	5.4%	16.2%	17.1%	21.7%	27.3%

Table 8

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.” The Early Development Network, Nebraska’s Part C Early Intervention Program, has identified 21 (64%) of the 33 infants with PHL still residing in Nebraska to be eligible for Early Intervention services. Services for 17 of the 21 infants were begun prior to 6 months of age and four were verified after 6 months of age. Three infants were not referred to EDN, four are pending verification as of 08/16/06, two are not eligible for services, and no information is available for three referrals. Twenty two (67%) of the infants diagnosed with a PHL have a medical home. Six (18%) have been enrolled in the Medically Handicapped Children’s Program, Nebraska’s Children with Special Health Care Needs Program, and eligibility is pending for three children with PHL.

Summary

- All the current birthing hospitals in Nebraska were conducting newborn hearing screening in 2005. All but one were conducting 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 has been met. In 2005, birthing hospitals reported screening the hearing of 99.4% of newborns.
- The overall refer rate of 3.4% for initial hearing screening during birth admission was within national norms during 2005.

- The rate of reported follow-up re-screening and/or diagnostic evaluation has continued to improve, increasing from 63% in 2001 to 84% in 2005.
- In 2005, follow-up re-screening occurred within one month of birth for 75.1% of those newborns for which follow-up activities were initiated. The average age at the time of the initiation of follow-up re-screening or diagnostic evaluation was 29.8 days.
- The average age at diagnosis of hearing loss was 122.4 days for those reported to NNHSP in 2005 and 47.1% of the evaluations occurred within 3 months of birth.
- The incidence of Permanent Hearing Loss identified (1.97 per thousand in 2005) and reported to NNHSP appears to be within the anticipated range of 1 to 3 per thousand.

ACTIVITIES – 2005

Funding

Health Resources Services Administration/Maternal and Child Health Bureau

The Nebraska Newborn Hearing Screening Program received \$125,000 in grant funds from the Health Resources Services Administration/Maternal and Child Health Bureau for the fifth consecutive year to fund the basic operations of the NNHSP. A \$30,000 supplemental grant was funded for Boys Town National Research Hospital to develop educational materials for health professionals and Spanish-speaking parents. The goals of the NNHSP are:

System Goal 1 – The hearing of all newborns in Nebraska will be screened during the birth admission or, if born out-of-hospital, by 1 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 6 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 – NNHSP will provide an effective structure for the newborn hearing screening and intervention system in Nebraska.

Centers for Disease Control and Prevention

The NNHSP also received \$145,850 from the Centers for Disease Control and Prevention for the first year of a three year Early Hearing Detection and Intervention Tracking, Surveillance and Integration cooperative agreement. The goals of the cooperative agreement are:

Goal 1 – Hearing screening results will be electronically reported to NNHSP for all occurrent births in Nebraska.

Goal 2 – Pediatric audiologic evaluations, medical evaluations, and developmental outcomes will be electronically reported to NNHSP for young children identified with a hearing loss.

Goal 3 – The NNHSP 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 NNHSP tracking, surveillance and integration project will be conducted and the results disseminated.

Advisory Committee

The NNHSP was developed based on the requirements identified in the Infant Hearing Act of 2000 and the protocols recommended 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 NNHSP 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 Nebraska Newborn Hearing Screening Program consists of 20 members representing medical, audiology, parents, family support, and education stakeholders (see Appendix B). The Advisory Committee met quarterly during 2005 and provided the following guidance to the NNHSP:

- Infants identified with hearing loss will be tracked for five years to document types of services received and outcomes.
- Criteria were developed to create a "Lost to Follow-up" category for instances when re-screening or diagnostic results are not accessible or available.
- The NNHSP will focus follow-up activities primarily for newborns born in Nebraska.
- The Advisory Committee provided input into the ongoing development of four

aspects of the NNHSP: 1) recommended follow-up and tracking protocols, 2) identification of the initial point of contact for parents of infants recently diagnosed with a hearing loss, 3) parent education materials (brochures, resource guides), and 4) hearing aid loaner bank feasibility.

Projects

Electronic Data System

Development of the Newborn Hearing Screening Module by QS Technologies for integration with the new HHS Electronic Vital Records System began in January, 2005, and the system was beta-tested in December, 2005. Development will continue during 2006 with implementation planned for late in the year. The integrated system will eliminate the need to manually record, transmit, and track demographic information on each newborn who “refers” or is discharged without a hearing screening and will increase the accuracy, consistency, and timeliness of newborn hearing screening information provided to the NNHSP by birthing hospitals.

Early Head Start ECHO Project

To begin the process of implementing periodic early childhood hearing screening in Nebraska, the ECHO project, developed by National Center for Hearing Assessment and Management and funded by the Head Start Bureau, trains Early Head Start programs to conduct OAE hearing screenings. A team consisting of four audiologists, an educator of the deaf, an early childhood training coordinator, and the American Academy of Pediatrics Early Hearing Detection and Intervention Nebraska Chapter Champion have been trained to conduct the ECHO trainings. OAE screening equipment is provided as part of this project. Two Early Head Start programs were trained to conduct OAE screenings with the infants and toddlers enrolled in their programs in November, 2004. Training of additional programs was temporarily halted due to funding delays from the Head Start Bureau. Discussions and technical assistance by phone and in-person occurred to overcome difficulties in following up with re-screenings and in timely submission of paperwork. Additional training was conducted to train new staff due to program restructuring and turnover.

Surveys

Pediatricians and family practice physicians in Nebraska received the Newborn and Infant Hearing Screening Survey developed by Boys Town National Research Hospital and National Center for Hearing Assessment and Management. Responses were received from 149 (17%) of the physicians; 35% of whom were pediatricians and 58% of whom were family practice physicians. Key findings from this survey were:

- 89% of physicians received newborn hearing screening results from the birthing facility.
- 89% responded that additional testing should be completed prior to 3 months of age.
- 22% responded that an infant could not be fitted with a hearing aid before 12 months of age.

- 12% responded that an infant could not be referred to Early Intervention before 12 months of age.

A second survey, the Professional Development Needs Assessment for newborn hearing screening coordinators in birthing facilities, was developed in collaboration with Boys Town National Research Hospital. Responses were received from 87% of the birthing facilities. Key findings from this survey are:

- 50% needed to occasionally train new staff or re-train current staff about hearing screening and 44% rarely needed to train new or current staff.
- 72% do not use a script to guide the discussion of hearing screening results with parents.
- Over 50% of the respondents indicated a need for training about these topics: supervisor modules, cultural competency, encouraging follow up, trouble-shooting, communicating with parents, and procedures.
- 82% indicated that web-based training would be helpful.

Revisions

Follow-up letters to Primary Care Providers and parents were simplified, parent resource guides were re-designed to include more recent and accessible materials about available services, the birthing facility reporting form was re-designed and simplified, the data tracking spreadsheet was revised to facilitate improved follow-up and reporting processes, and the monthly status report was expanded to provide more meaningful and comprehensive quality assurance information.

Educational Materials

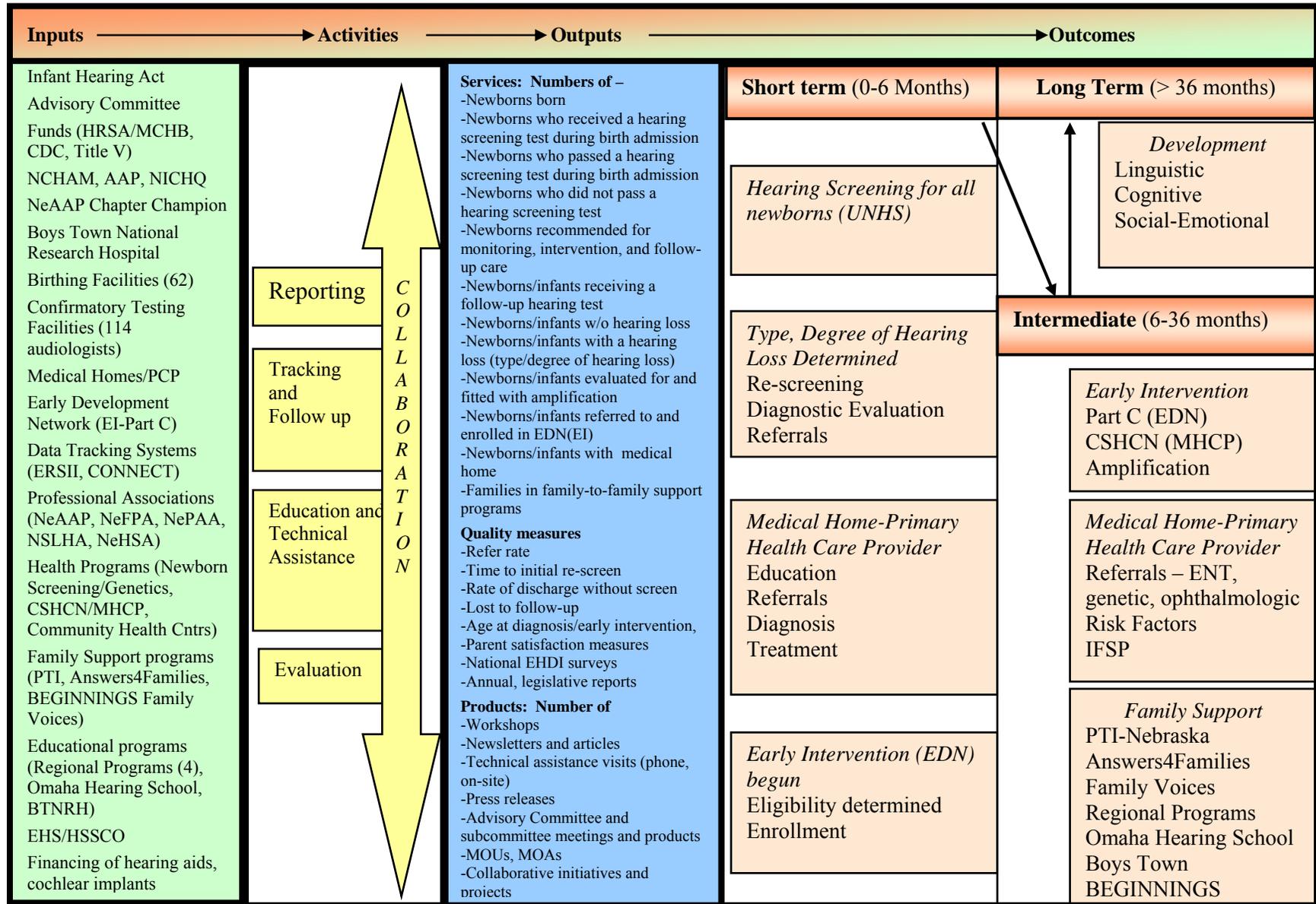
Bilingual parent education materials (brochures, videotapes) were provided at no charge to all birthing facilities. Preliminary work began on developing new parent education brochures and follow-up brochures written at an average health literacy level.

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 NNHSP program manager serves on workgroups to implement the following strategies:

- Develop and implement a collaborative initiative to promote the medical home approach as a standard of care for all children.
- Establish a comprehensive program to promote regular recommended pediatric visits for children, following the American Academy of Pediatrics and Bright Futures guidelines.
- Integrate parent to parent peer support systems into existing and new programs and services for families.
- Develop capacity for an early childhood data monitoring system through creation of an ECCS data agenda.

Nebraska Newborn Hearing Screening Program – Logic Model



Appendix B.

Advisory Committee Members

Committee Member	Group/Facility Represented
Steve Boney, PhD	Professor, Audiology University of Nebraska - Lincoln
Margaret A. Coleman	Nebraska Commission for the Deaf and Hard of Hearing
Lora Langley, RN, BSN	Ponca Tribe of Nebraska
Regina Watson, LPN-C	Hearing Screening Coordinator Tri County Area Hospital, Lexington
Mary Pat Moeller, PhD	Director, Center for Deafness Boys Town National Research Hospital
Stacie Ray, MS, CCC-A	Parent, Clinical Practice Supervisor – Audiology University of Nebraska - Lincoln
G. Bradley Schaefer, MD	Geneticist, Munroe-Meyer Institute, Nebraska Medical Center
Monica Seeland	Vice President, Nebraska Hospital Association
Britt Thedinger, MD	Otologist, Ear Specialists of Omaha
Donald M. Uzendoski, MD	Nebraska Chapter, American Academy of Pediatrics Early Hearing Detection and Intervention Chapter Champion
Robert Wergin, MD	Nebraska Academy of Family Physicians
Dawn Peters	Parent Training and Information-Nebraska
Eleanor Kirkland	Head Start State Collaboration Office (NDE)
Audrey Isaacson	Parent
Rhonda Fleischer	Liaison, Regional Programs for Deaf and Hard of Hearing Students (NDE)
Jeanne Garvin, MD	Medical Director Medically Handicapped Children's Program (CSHCN) (HHS)
Charlie Lewis	Answers4Families
Micaela Swigle	Co-Lead, Early Development Network (Part C) (HHS)
Julie Miller	State Genetics Coordinator, Newborn Screening Program (HHS)
Krystal Baumert	Follow-up Coordinator, Newborn Screening Program (HHS)
Jeff Hoffman, CCC-A	Manager, Newborn Hearing Screening Program (HHS)

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 disorders, 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 & NBHS) (402) 471-9731

Metabolic foods program, Patient Education materials translations and distribution

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Department of Health and Human Services Regulation and Licensure
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Pediatric Screening Laboratory Director, Joseph Quashnock, PhD (412) 220-2300 (Pennsylvania)

The staff of the **Nebraska Newborn Hearing Screening Program** are available to help with your questions at these numbers listed below. General areas of responsibilities are listed:

Jeffrey Hoffman, CCC-A, Newborn Hearing Screening Program Manager (402) 471-6770

Program planning, evaluation and management, systems development

Claire Covert, Staff Assistant (402) 471-3579

Follow-up, patient education materials distribution, data management, special projects

Mike Rooney, Administrative Assistant (402) 471-9731

Follow-up, patient education materials translations

Jim Beavers, Business Analyst (402) 471-1526

Business Analyst

(See address above)

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Hearing screening photos courtesy of Natus Medical, SonaMed Corp, National Center for Hearing Assessment and Management

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