

October, 2007

To persons interested in Newborn Screening:

You are receiving this Executive Summary of the Newborn Screening 2006 Annual Report as a stakeholder in the health of Nebraska's youngest citizens - our newborns. For the full report, access the Newborn Screening web-page at the Nebraska Department of Health and Human Services Web site at: <http://www.dhhs.ne.gov/nsp>.

This report covers the Newborn Screening System in Nebraska for the calendar year 2006.

The achievements that have occurred in Nebraska for both newborn blood-spot and hearing screening are due to the tremendous efforts of a "system" of people across the state. The State programs provide a framework, and to a degree, a "back-up" system via tracking and follow-up. We believe the most important work, however, occurs where people live, where they get their care, and where their babies are born.

This executive summary highlights a few of the many achievements and the continuing challenges of newborn blood-spot and newborn hearing screening systems in Nebraska. We hope you will find it interesting and useful.

Sincerely,



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Highlights from the 2006 Nebraska Newborn (Blood-spot) Screening Program, Annual Report:

- **In 2006, Nebraska added required screening for cystic fibrosis and congenital adrenal hyperplasia**, bringing the total number of conditions required to eight. The other required 6 are Biotinidase deficiency, Congenital Primary Hypothyroidism, Galactosemia, Hemoglobinopathies, MCAD and PKU.
- All newborns were offered supplemental screening at no extra cost and which required no extra blood. Supplemental screening included results from tandem mass spectrometry screening for about 26 additional fatty acid, organic acid and amino acid disorders.
- Supplemental screening was universally offered to all parents, but was not required. Parents must decide whether to consent to, or dissent from supplemental screening.
- **More than 97% of Nebraska parents consented to the supplemental screening.**
- The following babies were identified and entered into early treatment and intervention, most with specialty clinics specific to their type of condition:
 - 4 babies with partial biotinidase deficiency (treated)
 - 1 baby with congenital adrenal hyperplasia
 - 10 babies with congenital primary hypothyroidism
 - 13 babies with cystic fibrosis
 - 6 babies with hemoglobinopathies
 - * (3 with Sickle Cell disease, 2 Sickle Hgb. C disease and 1 hemoglobin-E disease)
 - 4 babies with hyperphenylalaninemia requiring treatment
 - 1 baby with 3-MCC Deficiency (3-methylcrotonyl-CoA Carboxylase Deficiency)
 - 1 baby with IBCD (Isobutyryl-CoA Dehydrogenase Deficiency)
 - 2 babies with transient tyrosinemia
 - 1 with presumed cobalamin defect

In 2006 newborn screening efforts resulted in successfully identifying and treating 43 newborns affected with conditions in time to prevent mental retardation, other chronic illness and disability, and infant death.

Individually each condition is quite rare. However in 2006, 1 in 624 babies were identified through newborn screening to have one of the clinically significant screened conditions!

- Overall in 2006, **97.32% of parents consented to the supplemental newborn screening panel** from tandem mass spectrometry (MS/MS).

- In 2006 the March of Dimes Nebraska Chapter met with representatives of the Nebraska Health & Human Services System to promote mandatory screening of all conditions detected on the MS/MS.
- Nebraska contracts for Newborn Screening Laboratory Testing Services, with Pediatric Screening Laboratory in Pennsylvania. Courier service to ensure overnight specimen delivery is an important element of that contract. Monitoring of turn around times becomes even more critical when sending specimens out of state:
- Ensuring a quality specimen is collected and handled correctly the first time is important to ensuring rapid identification of newborns affected with screened conditions. Unsatisfactory specimens, specimens collected too early (at less than 24 hours of age), and specimens collected post transfusion can create delays in the timely identification of newborns. The Newborn Screening Advisory Committee monitors these measures each quarter:
 - 282 or 1.04% of all newborns had a specimen collected before 24 hours. Babies to be transferred accounted for 44.45% of these, babies to be transfused accounted for 10.7% of these. The reason for early collection is unknown for the remaining 44.85%.
 - 100 or 0.38% of all newborns initial specimens (26,819) were "unsatisfactory"
 - The mean-average turn around time from birth to results ranged between 5.2 - 5.45 days.
- The Newborn Screening Advisory Committee began deliberations to address financial sustainability for this important public health program in light of federal funding cuts that have occurred in the Title V Maternal and Child Health Block Grant which helps support Nebraska's Newborn Screening System.

Highlights from the 2006 Nebraska Newborn Hearing Screening Program Annual Report:

Follow-up services occurred within one month of birth for 77% of the newborns for whom follow-up activities were initiated. The average age at initiation of follow-up re-screening or diagnostic evaluation was 27.4 days.

The Infant Hearing Act of 2000 established newborn hearing screening in Nebraska. Birthing facilities are to educate parents about hearing screening, to include newborn hearing screening as part of the standard of care, and, by December 1, 2003, to conduct

The hearing of 98.9% of newborns was screened during the birth admission.

hearing screenings for at least 95% of the newborns born in Nebraska.

- All of the current birthing facilities conducted newborn hearing screening in

2006.

- The refer rate for hearing screening during birth admission was 3.8%.

- The reporting rate of completed follow-up re-screening and/or diagnostic evaluation has continued to improve, increasing from 63% in 2001 to 87% in 2006.
- The average age at diagnosis of hearing loss was 55 days for those reported to NNHSP in 2006.
- Early Intervention services were initiated for 62% of the infants diagnosed with permanent hearing loss. Over 85% of those were verified prior to 6 months of age.

In 2006, 26 infants with permanent hearing loss were identified, an incidence of 1:1000.

Funding - The Nebraska Newborn Hearing Screening Program received grant funds from the Health Resources Services Administration/Maternal and Child Health Bureau to fund the basic operations of the NNHSP. The NNHSP also received funding from the Centers for Disease Control and Prevention for the second year of a three year Early Hearing Detection and Intervention Tracking, Surveillance and Integration cooperative agreement.

Advisory Committee - The Advisory Committee of the Nebraska Newborn Hearing Screening Program consists of 22 members representing medical, audiology, parents, family support, and education stakeholders. The Advisory Committee met quarterly.

Electronic Data System - Development of the Newborn Hearing Screening Module by QS Technologies/Netsmart for integration with the Electronic Vital Records System continued during the first ten months of 2006. To meet the target implementation date of January 1, 2007, selected staff of the largest birthing facilities were trained in November and December, with the rest of the birthing facilities scheduled for training in January and February, 2007. The integrated system is based on the birth records and provides for the reporting of hearing screening results for all occurrent births in Nebraska. This will eliminate the need to manually record, transmit, and track demographic information on each newborn who “refers” or is discharged without a hearing screening.

Educational Materials - The new parent education brochure “Can Your Baby Hear?” and the new follow-up brochure “Your Baby Needs Another Hearing Screening” were written at an average health literacy level and are now available in ten languages (English, Spanish, Arabic, Russian, Vietnamese, Chinese, French, Nuer, Dinka, and Anuak). The Parent Resource Guide was updated with new materials about educational and health services, unbiased information about communication options, and a Funding Toolkit offering financial assistance information to parents of children diagnosed with hearing loss.

National Initiative for Child Healthcare Quality Learning Collaborative —Nebraska was one of eight states selected to participate in a Learning Collaborative, funded by the Health Resources Services Administration/Maternal and Child Health Bureau and developed by NICHQ. The purpose of the project is to reduce the number of babies who are lost to follow-up by developing strategies that are shown to be effective through small tests of change.

Hearing Aid Loaner Bank— A joint partnership between the University of Nebraska – Lincoln and the NNHSP was established to study the feasibility of establishing a hearing aid loaner bank in Nebraska for young children identified with a Permanent Hearing Loss. Two audiology graduate students at UNL explored the funding, administration, and selection criteria for states with hearing aid loaner banks, the capacity of audiologists licensed in Nebraska to fit hearing aids for newborns and infants, hearing aid manufacturers’ programs to provide discounts and support for hearing aid loaner banks, funding sources through private and non-profit organizations, and cost estimates to develop and maintain a hearing aid loaner bank.

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EXECUTIVE SUMMARY OF NEBRASKA'S 2006 ANNUAL REPORT ON NEWBORN SCREENING
INCLUDING
SCREENING FOR METABOLIC AND INHERITED DISORDERS
AND
NEWBORN HEARING SCREENING

For full report see:
Web-site at <http://www.dhhs.ne.gov/nsp/>
or contact the Newborn Screening Program at 402 471-9731.



NEWBORN SCREENING IN NEBRASKA

**Newborn Screening for Metabolic and
Inherited Disorders**

AND

Newborn Hearing Screening



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