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UNIVERSAL NEWBORN HEARING SCREENING IN ILLINOIS 2003 – 2004

August 2006

**UNIVERSAL NEWBORN
HEARING SCREENING
IN ILLINOIS
2003 – 2004**

Illinois Department of Public Health
Division of Epidemiologic Studies

August 2006

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2003-2004

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GLOSSARY

Audiologist is a person licensed by the state of Illinois' Department of Financial and Professional Regulation to provide audiological services.

Automated auditory brainstem response (AABR) is an objective electrophysiologic measurement of the brainstem's response to acoustic stimulation of the ear, obtained with equipment that automatically provides a pass/refer outcome.

Diagnostic audiological evaluation is the physiologic and behavioral procedures required to evaluate and diagnose hearing status.

Early Intervention is a statewide, family-centered service system to find and help children younger than 36 months who have disabilities or developmental delays. These infants and toddlers are eligible through the Illinois Early Intervention Services System for special early intervention services defined in Part C of the Individuals With Disabilities Act (IDEA).

Hospital means, in the setting of newborn hearing screening, a facility licensed by the state of Illinois under the Hospital Licensing Act providing obstetrical and neonatal services.

Medical diagnostic evaluation is the examination and medical procedures provided by an otolaryngologist, otologist or other qualified personnel to evaluate otologic status.

Otoacoustic emissions testing is a specific test method that elicits a physiologic response from the outer hair cells in the cochlea.

Otolaryngologist is a physician trained in the medical and surgical management and treatment of patients with diseases and disorders of the ear, nose, throat, and related structures of the head and neck.

Otologist is a physician who specializes in treatment of the ear.

Screening is the completion of one or more objective, physiologic, electronic tests administered to determine the need for further diagnostic testing by an audiologist and physician. The screening shall be performed by individuals who have been appropriately trained in the procedure and instrumentation used by the hospital.

Significant hearing loss is a dysfunction of the auditory system of any type or degree that is sufficient to interfere with the acquisition of speech and language skills.

INTRODUCTION

Significant hearing loss is one of the most common major abnormalities present at birth. About one in every 1,000 children is born profoundly deaf [National Institutes of Health (NIH) Consensus Statement]; four times as many are born with moderate or severe bilateral hearing loss (Task Force on Newborn and Infant Hearing). Infants in neonatal intensive care units are 10 to 20 times more likely to have significant hearing loss than the healthy population (Task Force on Newborn and Infant Hearing).

Prior to the 1993 NIH recommendation of universal newborn hearing screening, the average age of detecting hearing impairment in infants and toddlers was 3 years. The Task Force on Newborn and Infant Hearing referred to a survey, published in 1998, that found that the median age of detection of severe to profound hearing loss in the United States was 13 months. Mild to moderate hearing loss was not identified until a median age of 17 months.

The first three years of life are the most important for language and speech development. Consequently, for many infants and young children with unidentified hearing impairment, much of the crucial period for language and speech development may be lost. This in turn leads to lower reading abilities, poorer academic achievement and fewer career opportunities (Task Force on Newborn and Infant Hearing).

According to current estimates, permanent congenital hearing loss of greater than 25 dB HL (hearing level) in the poorer ear is present in at least three infants per thousand (White 1997). Given Illinois birth rates (182,393/2003; 176,272/2004), it would be expected that approximately 528 -547 infants would be born each year with significant, congenital hearing loss. Research conducted by Yoshinaga-Itano (1995) indicates that children identified with hearing loss who are linked to appropriate intervention by 6 months of age can develop normal language skills.

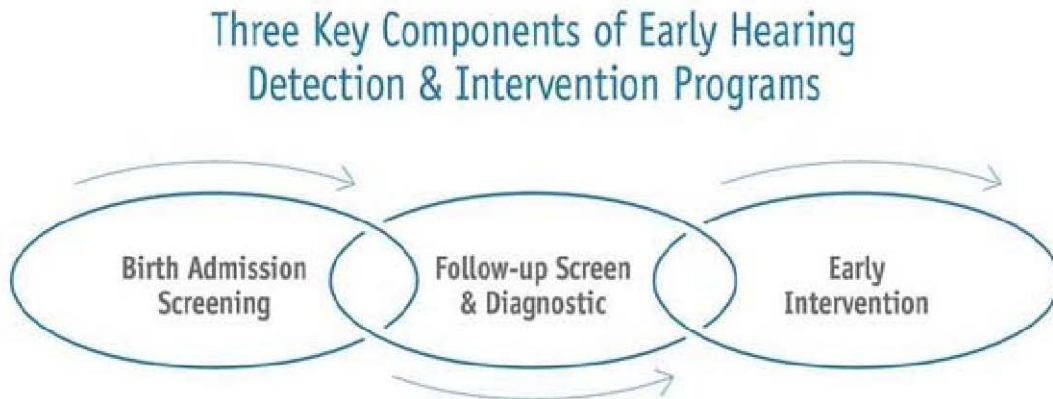
A child's hearing impairment should, therefore, be identified as early in life as possible, so that he or she can receive timely and appropriate interventions. The interventions will then take full advantage of the plasticity of the child's developing nervous system, optimizing his or her social, emotional, psychological and academic development.

In 1993, the NIH recommended that all children be screened for hearing loss by 3 months of age and that objective tests be used rather than behavioral testing or screening only those children identified as being at high risk. In 1994, the Joint Committee on Infant Hearing (JCIH) endorsed this recommendation and suggested that screening should take place before a newborn was discharged from the birth hospital to ensure that most children would be screened. In July 1999, Illinois passed the Hearing Screening for Newborns Act (410 ILCS 213/1), which legislatively mandated universal newborn hearing screening to become effective in Illinois on December 31, 2002.

The JCIH comprehensive and widely peer-reviewed position statement in 2000 again recommended universal screening of hearing loss before hospital discharge and went on

to identify principles and guidelines for birth hospital and state level programs. The JCIH 2000 statement promoted a system composed of screening before hospital discharge, follow-up and diagnosis for infants needing follow-up and the intervention and habilitation for infants identified with hearing loss (Figure 1.)

Figure 1



Some hearing impairment will develop during the first years of life, and will not be present at birth. It is important, therefore, to continue to watch for signs of changes in hearing status as children grow.

Early indications show that the state's newborn hearing program is effective in identifying children with congenital hearing losses at an earlier age. Prior to universal newborn hearing screening implementation, a 1995 study by Kittrell and Arjmand revealed that in Illinois the average age of identification of hearing loss was 20.2 months and the average age of initial amplification was 31.7 months. By the end of calendar year 2004, two years after universal newborn hearing screening implementation, the average age of diagnosis of infants reported to the Illinois Department of Public Health (IDPH) had greatly improved to 3.8 months of age.

METHODS

THE ILLINOIS NEWBORN HEARING PROGRAM

Universal newborn screening for hearing loss began in Illinois on January 1, 2003. The goals of the Illinois Newborn Hearing Program are universal hearing screening and detection of hearing loss in infants before 3 months of age, and the initiation of appropriate intervention no later than 6 months of age. To this end, IDPH is charged with maintaining a registry of infants in need of follow-up. In addition, IDPH is required to refer children with confirmed hearing losses for services to minimize problems caused by the impairment. For further information about the Illinois Newborn Hearing Program, contact the IDPH Newborn Hearing Program team at (voice) 217-782-4733 or (TTY) 800-547-0466.

The Illinois Newborn Hearing Screening Program maintains the registry of infants in need of follow-up testing through three stages:

- screening;
- audiologic evaluation of those with an abnormal result (confirmation); and
- early intervention for those with confirmed hearing impairment.

SCREENING

Since 2003, almost all infants born in Illinois hospitals have been screened (in both ears) for congenital and neonatal-onset hearing loss prior to the discharge from their birth hospitals. These babies include infants served in both the well-baby nurseries and the neonatal intensive care units. Babies born in alternative birthing facilities, including those born at home, are recommended to be screened before 1 month of age.

In certain circumstances, babies may not be screened. Parents have the right to refuse the newborn hearing screening when it is in conflict with their religious beliefs or practices (410 ILCS 213/25). Otherwise, a small number of infants may not be screened including:

- those who die before the hearing screening is performed;
- those discharged before screening;
- those who were transferred to other hospitals before being screened; and
- those born outside a hospital.

The Illinois Newborn Hearing Program aims to detect hearing impairment including permanent bilateral or unilateral, sensory or conductive hearing loss, averaging 30 to 40 dB or more in the frequency region 500 through 4000 Hz. Hearing impairment in this range will most impact speech acquisition.

Screening Methods

The American Academy of Pediatrics and Joint Committee on Infant Hearing recommend two methods of screening: AABR- automated auditory brainstem response or EOAE – evoked otoacoustic emissions. Illinois hospitals use either otoacoustic

emissions (OAE) or automated auditory brainstem response, alone or in combination, depending on the chosen screening equipment. Hospitals are free to select their own brand of equipment provided it meets the technological parameters addressed in the rules. The screening equipment must be designed for newborn hearing screening, measure a physiologic response, measure the status of the peripheral auditory system, be highly correlated with hearing status and be implemented with objective response criteria. Both AABR and OAE screening methodologies meet the rule's standards. The methodology also must detect unilateral or bilateral hearing loss greater than or equal to 35dBHL (a false negative rate of zero) and have a false positive rate of 3 percent or less.

Both hearing screening methods are objective and physiologic measures that do not need the active patient response required in traditional audiologic evaluation tests. The tests are painless, noninvasive, and easy to perform. Usually the tests are completed within a few minutes.

Otoacoustic emissions

An otoacoustic emission is a weak "echo" type sound emitted by the ear soon after a sound is perceived. The OAE measures the stimulated acoustic energy generated in the cochlea (inner ear) that travels through the middle ear into the ear canal where it is sensed with a miniature microphone.

To perform the OAE, a tiny, flexible plug is inserted into the baby's ear. Specific sounds are generated through the plug. A miniature microphone in the plug records the otoacoustic responses of the inner ear in reaction to the transmitted sounds. The test is usually done while the baby sleeps.

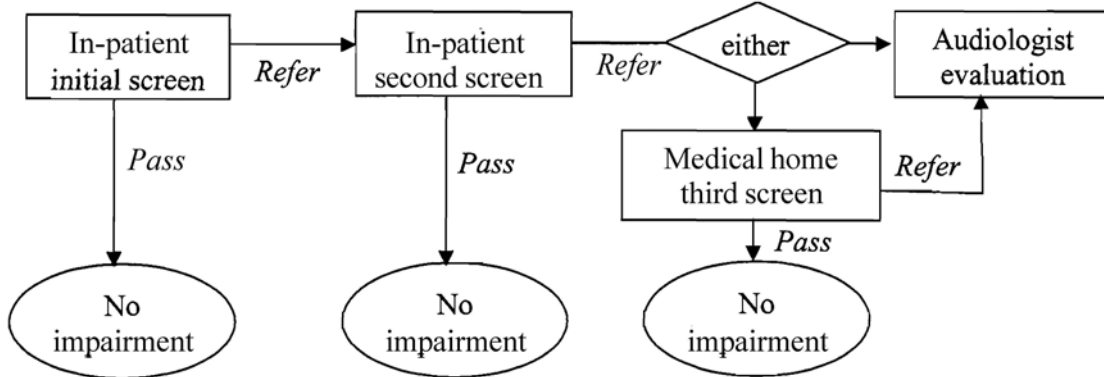
Automated auditory brainstem response

The AABR provides a complete screen of the auditory pathway up to the brainstem (including the middle ear, the inner ear, and the VIII nerve). When an AABR is performed, electrodes are placed on the forehead, nape of the neck and shoulder (ground). With AABR screening, a click stimulus at one loudness level is presented to each of the child's ears. The child's response is compared to a template of children with normal hearing. If the responses match, the child passes the screening; if the response does not match, then the child is referred for further testing. Screening by AABR can be completed immediately after birth and a stringent statistical pass criterion is employed that eliminates bias from interpretation and user skill level. AABR is a screening tool used for infant who have reached at least 34 weeks conceptional age until the child turns 6 months of age.

A review by Clemens and Davis reported that the false-positive rates of early (prior to 1999) UNHS programs ranged between 2.5 and 8 percent. Higher false-positive rates may lead to a variety of unnecessary negative effects, including emotional trauma, possible disease labeling, and increased expense in terms of time and money. Clemens and Davis (2001) did follow-up research and demonstrated the efficacy of rescreening all infants who were referred on their initial hearing screen before hospital discharge. Rescreening prior to discharge reduced the false-positive rate to a low rate of 0.8 percent.

Illinois law mandates that every baby whose initial screen indicates a possible hearing problem in one or both ears will be screened a second time prior to discharge (Figure 2).

Figure 2. Flow chart of the Illinois two-step newborn hearing screening method



If a baby is transferred without written documentation of a completed hearing screening, the hearing screening should be completed by the receiving hospital, prior to discharge.

A referral after two in-patient screens does not necessarily indicate that the infant has a congenital or permanent hearing loss. The referrals may have been due to transient fluid or vernix in the canal that has cleared by the time of the outpatient screen or test. The third screening or recommended diagnostic evaluation is performed after discharge on an outpatient basis; preferably well within 3 months of age.

Role of the Newborn Hearing Program

In addition to tracking the data reported to IDPH, the Newborn Hearing Program makes substantial efforts to obtain out-patient audiological results for every child referred for further testing. These efforts include letters to parents and physicians; telephone calls to hospitals; and referrals to local health departments. Those families who refuse the initial screening must do so based on religious beliefs, and in writing. The hospital provides the parents with information on the importance of hearing screening; these families also receive letters from IDPH.

EVALUATION AND INTERVENTION

Babies who do not pass the two-step hearing screen or any subsequent re-screening are tracked to make sure they receive a follow-up diagnostic evaluation (Figure 2) to confirm the existence of (and determine the type and severity) or rule out hearing impairment. Ideally, this evaluation takes place before the baby is 3 months old. If a child has been referred to an audiologist, but no results received, the Newborn Hearing Program will

contact parents, physicians, hospitals, audiologists and local health departments to assure that hearing impairment is either confirmed or ruled out.

The Joint Committee on Infant Hearing recommended a battery of tests for children referred to an audiologist for diagnostic evaluation (Table 1).

Table 1. The Joint Committee on Infant Hearing's recommended test battery for children referred to an audiologist for evaluation.

Test	Age at evaluation	
	Birth to 6 months	6 to 36 months
Child and family history	✓	✓
Electrophysiologic measure of threshold, using frequency-specific stimuli	✓	✓
Speech detection and recognition measures		✓
Behavioral response audiometry		✓
Parental report of emerging communication and auditory behaviors	✓	✓
Screening of the infant's communication milestones		✓

If an audiologist determines that a child's hearing is impaired, the family should be referred for a medical evaluation and to appropriate intervention programs.

Medical Evaluation

After the confirmation of hearing impairment, a child should be evaluated to identify the etiology of his or her hearing loss. This will include a family history, clinical history, physical examination, laboratory, radiologic studies, and perhaps a genetic consultation. Once the reason for the hearing loss is determined, the most appropriate treatment and interventions can be decided.

Early Intervention

Any number of professionals may be involved in the services offered to a child and his or her family including:

- the audiologist and audiological team;
- primary care physician;
- speech pathologists;
- educators of children who are hard of hearing or deaf;
- language and communication development specialists;
- early intervention specialists;
- otolaryngologists;
- plastic surgeons;
- child development specialists;
- deaf mentors; and
- alternative language (cued speech or sign language) teachers.

DATA AND REPORTING

(IDPH's Vision and Hearing Program, located within the Division of Health Assessment and Screening, administers the Illinois Newborn Hearing Program in conjunction with the Illinois Department of Human Services, the Division of Specialized Care for Children (DSCC), and the state's Children with Special Health Care Needs (CSHCN) Program. By law, all hospitals are required to conduct physiological hearing screens on all infants delivered under their care and to notify the IDPH Newborn Hearing Program of the test results on all infants. Hearing screening is completed using a non-invasive technology that, within a few hours of birth, can differentiate those infants with no hearing loss from those who require further testing.

The Newborn Hearing Program staff has successfully trained all 139 birthing hospitals in Illinois to report their local newborn hearing screening data in an encrypted electronic format. Electronic reporting and data management is done using a central database and 139 local applications of the Hi*Track Data Management System, the software system created by the National Center for Hearing Assessment and Management (NCHAM) at Utah State University, for newborn hearing screening data management and tracking. Hospitals are required to send their screening data to IDPH weekly by encrypted email attachment. This allows for the timely submission of screening and follow-up data and a means of monitoring receipt of data by the state.

At the state level, Hi*Track is used for centralized, statewide program management of tracking and follow-up activities. Upon receipt of the data, the files are merged into the central Hi*Track database from which letters to physicians and parents are generated, monitoring of tracking activities takes place, and data management, report generation and program evaluation can be done.

Annual prevalence is defined as

$$10,000 \times \frac{\text{number of infants with hearing impairment in a given year}}{\text{number of children screened in the same year}}$$

Calculation of Rates and Confidence Intervals

Occurrence of confirmed congenital hearing impairment is a rare event; occurrences follow a Poisson distribution. Exact confidence intervals were used (Armitage and Berry, 1987, page 134). Where there are a large number of cases of hearing impairment, the confidence interval is narrow, indicating that the rate is stable. Where there are few such cases, the confidence interval becomes very wide, indicating that the rate is not very stable and a small change in the number of infants born with hearing impairment could result in a large change in the rate.

If we assume that the rate of the congenital hearing impairment is constant over time, then we may consider the observed rate to be a sample from the underlying distribution over time. In this context, we can calculate a confidence interval for a rate. Our

observed sample for a given time period is an independent sample drawn from the Illinois newborn infant population. If this is repeated many times, and a confidence interval is calculated every time, then 95 percent of these confidence intervals will cover the unknown, true Illinois population incidence of congenital hearing loss.

To compare two rates, it is important to look not just at their values, but also their confidence intervals. As a conservative approximation, if two confidence intervals overlap, then there is no evidence that the two rates are really different. If two confidence intervals do not overlap, then the rates are said to be statistically different. In this report, 95 percent confidence intervals are used; where the confidence intervals do not overlap the rates are statistically different at the 5 percent level ($p < 0.05$).

RESULTS

Screening

The Illinois Newborn Hearing Program established the statewide tracking system in December 2002 and started tracking its first referral case of congenital hearing impairment in 2003. Hearing screening results were reported for 89.6 percent of the live births in 2003 and for 95.2 percent of the live births in 2004. This increase shows the great improvements in obtaining reporting compliance during 2003. Table 2 shows screening compliance as reported to IDPH. The second row (*Any screening*) indicates children who were screened at least once. The third row (*Completed screening*) indicates children who either passed a screening or were referred to an audiologist after a screening. Table 2 also shows the improvements made in 2004, with a higher percentage of children completing the screening process, and lower percentages of cases with no information.

Table 2. Screening compliance during the first two years of the Illinois Newborn Hearing Program

Children	2003		2004	
	Number	Percent	Number	Percent
Reported	163,431		171,981	
Any screening (in- or out-patient)	161,151	98.6	170,712	99.3
Completed screening	158,667	97.1	168,558	98.0
Deceased	552	0.3	626	0.4
Screening not complete	4,212	2.6	2,767	1.6
Refused	49	1.2	27	1.0
Pending screening	77	1.8	211	7.6
Inconclusive	30	0.7	21	0.8
Lost to follow-up	305	7.2	304	11.0
Trying to locate/duplicate	3,750	89.0	2,204	79.6

Source: Illinois Department of Public Health Newborn Hearing Program HI*TRACK 3.5 Flow Chart as of 12/02/2005

In 2003, 158,347 (98.3 percent) of those with any screening were screened before the recommended 1 month of age (or discharge from hospital). In 2004, 167,939 (98.4 percent) of those with any screening were screened by 1 month of age (or discharge).

Referred for Evaluation

Among those whose screening was completed, 1,053 (6.6/1,000 screenings) children were referred for further evaluation in 2003. A similar number (1,082 or 6.4/1,000 screenings) were referred in 2004. A referral may have occurred because the screening results indicated a possible problem, or because a parent or physician was concerned about a child’s hearing for some other reason. Table 3 shows the rate of confirmed hearing loss in Illinois infants.

Table 3. Rate of confirmed hearing loss in Illinois infants as reported to IDPH.

	2003	2004
Number of children with completed screening	158,667	168,558
Number of infants with confirmed hearing loss as reported to IDPH	103	130
Rate of confirmed hearing loss/10,000 completed screens	6.5	7.7
95% Confidence Interval	(5.3, 7.9)	(6.4, 9.2)

Source: Illinois Department of Public Health Newborn Hearing Program HI*TRACK 3.4 State EHDI report for CDC as of 12/2/2005

Table 4 shows the degree of hearing impairment among those who have confirmed hearing impairment. Two-thirds of those with confirmed hearing loss have loss in both ears. In about half the children, their hearing loss is severe or profound.

Table 4. Degree of hearing impairment, among those reported with confirmed hearing loss.

	2003		2004	
	Number	Percent	Number	Percent
Confirmed Hearing Loss	103		130	
Bilateral	67	65.0	93	71.5
Mild or Moderate	34	50.7	44	47.3
Severe	15	22.4	24	25.8
Profound	18	26.9	25	26.9
Unilateral	36	35.0	37	28.5
Mild or Moderate	18	50.0	20	54.1
Severe	10	27.8	13	35.1
Profound	8	22.2	4	10.8

Source: Illinois Department of Public Health Newborn Hearing Program HI*TRACK 3.4 State EHDI report for CDC as of 12/2/2005

In 2003, the median age for identification of hearing loss was 3.5 months; in 2004 it was 3.9 months. Thus by 2004, just under half the children with confirmed hearing loss were diagnosed by the recommended 3 months of age.

CONCLUSION

Prior to universal newborn hearing screening implementation in Illinois, the average age of identification of hearing loss was 20.2 months. By the end of calendar year 2004, two years after universal newborn hearing screening implementation, the average age of diagnosis of infants with permanent, congenital hearing loss reported to Public Health was 3.8 months of age. Early identification and intervention allow deaf and hard of hearing children the opportunity to develop language skills during a period of neural plasticity that would otherwise be forfeited, banishing them into a world of social isolation and educational malaise. As more children are identified at earlier ages and intervention begun in a timely manner, society will reap the economic benefits of reduced special education costs and increased potential for academic achievement and gainful employment. A price tag cannot be put on the benefits to families and their ability to interact with deaf and hard of hearing children throughout the child's lifetime.

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