

# The Status of Diagnostic Testing following Referral from Universal Newborn Hearing Screening

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## Abstract

The Joint Committee on Infant Hearing 2000 position statement includes guidelines for the development of Early Hearing Detection and Intervention programs. These guidelines provide specific recommendations for the audiologic test battery for infants who fail a newborn infant hearing screening. The recommended test battery includes electrophysiologic measures such as the ABR, frequency specific electrophysiologic tests, bone-conducted ABR, OAEs, tympanometry using high frequency probe stimuli, and acoustic reflexes. In the Commonwealth of Kentucky, 42 centers are listed as providing follow-up diagnostic testing services for infants failing the newborn hearing screening. The purpose of this investigation was to determine how many of these centers were abiding by the Joint Committee guidelines. Results show that only three of 42 centers listed are providing services that meet the guidelines. Less than 50% of infants identified with hearing loss are referred for genetic evaluations by the audiologist. Only 19 of the 42 sites listed provide amplification services for infants identified with hearing loss.

**Key Words:** Auditory evoked potentials, diseases, hearing disorders, infant, neonatal screening, newborn

**Abbreviations:** ABR = auditory brainstem response; ASSR = auditory steady state response; CSHCN = Commission for Children with Special Health Care Needs; DPOAE = distortion product otoacoustic emissions; EHDI = Early Hearing Detection and Intervention; JCIH = Joint Committee on Infant Hearing; KISS = Kentucky Infant's Sound Start; NIH = National Institutes of Health; OAE = otoacoustic emissions; TEOAE = transient evoked otoacoustic emissions; UNHS = universal newborn hearing screening

## Sumario

El comunicado del año 2000 del Comité Conjunto sobre Audición Infantil incluye guías para el desarrollo de programas de Detección e Intervención Temprana Auditiva. Estas guías aportan recomendaciones específicas sobre las baterías de pruebas audiológicas para aquellos infantes que fallan un tamizaje auditivo neonatal. La batería de pruebas recomendada incluye mediciones electrofisiológicas tales como un ABR, pruebas electrofisiológicas con especificidad de frecuencia, ABR transmitido por vía ósea, OAE, timpanometría utilizando estímulos de alta frecuencia, y reflejos acústicos. En la Comunidad de Kentucky se citan 42 centros capaces de brindar servicios de evaluación diagnóstica de seguimiento para bebés que fallan el tamizaje auditivo neonatal. El propósito de esta investigación fue determinar cuántos de estos centros seguían las guías del Comité Conjunto. Los resultados muestran que sólo tres de los 42 centros en la lista brindan servicios que cumplen con estas guías.

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Menos del 50% de los niños identificados con trastornos auditivos son referidos por el audiólogo para evaluaciones genéticas. Sólo 19 de los 42 sitios en la lista brindan servicios de amplificación para los niños identificados con alteraciones auditivas.

**Palabras Clave:** Potenciales evocados auditivos, enfermedades, trastornos auditivos, infante, tamizaje neonatal, recién nacido

**Abreviaturas:** ABR = Respuesta auditiva del tallo cerebral; ASSR = Respuesta auditiva de estado estable; CSHCN = Comisión para Niños con Necesidades Especiales de Salud; DPOAE = emisiones otoacústicas por productos de distorsión; EHDI = Detección e Intervención Auditiva Temprana; JCIH = Comité Conjunto sobre Audición Infantil; KISS = Programa de Iniciación Sonora Infantil de Kentucky; NIH = Institutos Nacionales de Salud; OAE = emisiones otoacústicas; TEOAE = emisiones otoacústicas por transitorios; UNHS = Tamizaje Auditivo Neonatal Universal

The National Institutes of Health (NIH) Consensus Statement (1993) noted that the average age of identification of infants and children with hearing loss was approximately three years in the absence of hearing screening programs at birth. The NIH therefore recommended the goal of universal screening for hearing impairment by three months of age. As is well known, late identification of hearing loss results in the child missing out on critical developmental time for speech and language, auditory system development, social, emotional, cognitive, and academic development, as well as the eventual consequences associated with reduced vocational and economic potential. The NIH report also noted that approximately 50% of the children with hearing loss were being missed using the more traditional high-risk registry as a means to identify infants who would be screened.

Universal newborn hearing screening (UNHS) is commonly understood to be a program whereby all newborns would have a hearing screening prior to discharge from the hospital. Conceptually the birth screening is only the initial component of a much more complex program that includes referral for follow-up testing; diagnostic testing procedures used to determine the presence of, type, degree, and configuration of hearing loss; decisions determining appropriate intervention for the hearing loss; and implementation of treatment plans. In addition, the program for determination of hearing loss at birth would include defining the roles and responsibilities of the various professionals involved with the process.

The principles and guidelines for establishing a universal newborn hearing screening program were provided by the Joint Committee on Infant Hearing (JCIH) *Year 2000 Position Statement* (Joint Committee on Infant Hearing et al, 2000). The JCIH position statement includes guidelines for development of Early Hearing Detection and Intervention (EHDI) programs, with the goal of maximizing linguistic and communicative competence and literacy development for children who are hearing impaired or deaf. These guidelines expanded on the NIH recommendations to not just screen for hearing loss by three months but to identify the degree, type, and configuration of the loss by three months of age, with intervention by six months of age, and audiological and medical monitoring for children with risk indicators for delayed onset or progressive hearing loss up to three years of age. In addition, the guidelines include the necessity to establish appropriate early intervention programs. In order to meet the broad goals of the position statement, the implementation of successful newborn hearing screening programs across the country was, and continues to be, an important first step in the process.

#### DIAGNOSTIC TESTING OF INFANTS FAILING THE SCREENING

Over the past decade, the development and implementation of newborn hearing screening programs has received significant attention. The accepted goal of screening by

the age of three months appears to be embraced by most professionals and organizations. The technology and methodology of assuring that all newborns are screened have continuously been evaluated and improved. The first step in the process of identification and treatment of hearing loss through universal newborn hearing screening programs is progressing on a local, state, and national level.

Equally important to the success of a universal newborn hearing screening program is the accessibility to appropriate diagnostic testing for the detection and characterization of hearing loss. As previously noted, the JCIH position statement recommends identification of hearing loss by three months of age and also includes a protocol for the evaluation of hearing in newborns referred after failing a hearing screening.

The JCIH recommended a test battery for evaluation of infants failing the newborn screening that includes both physiologic measures and developmentally appropriate behavioral tests. The stated purpose of the recommended test battery is to assess the integrity of the auditory system and to obtain ear specific estimates of type, degree, and configuration of hearing loss. This same information can also be used to develop appropriate intervention options.

The JCIH guidelines recommend that the test battery begin with a child and family history. For infants birth to six months of age, the test battery must include electrophysiologic measures such as the auditory brainstem response (ABR) to measure threshold or other tests using frequency-specific stimuli such as tone bursts. Bone-conducted ABR should be performed when necessary. The assessment must include otoacoustic emissions (OAEs), measures of middle ear function (tympanometry) using the appropriate high-frequency probe stimuli (necessary for infants under six months of age), and acoustic reflex thresholds. Observation of the infant's behavioral response to sound should be conducted, and parental report of emerging communication and auditory behaviors should be requested.

For infants and toddlers 6 through 36 months of age, behavioral response audiometry using visual reinforcement or conditioned play audiometry should be chosen according to the child's developmental age.

The battery should also include OAEs, acoustic immittance measures including acoustic reflex thresholds, and speech detection and recognition measures. A parental report regarding auditory and visual behaviors, and a communication screening, should be obtained. The recommendations also state that electrophysiologic tests, such as ABR, should be performed at least one time to confirm type, degree, and configuration of hearing status.

The JCIH guidelines also provide general recommendations regarding amplification and audiologic habilitation. The majority of infants and children identified with hearing loss will benefit from some form of amplification. The chosen device should be fit as soon as possible following identification. According to the JCIH document, infants and toddlers with confirmed hearing loss should be referred to the appropriate physicians, including the primary care physician (medical home) and an otolaryngologist, as well as any others that may be indicated on an individual basis. Referral to state early intervention programs is also recommended. Appropriate audiologic intervention should begin as soon as possible, preferably prior to six months of age or within one month of identification.

In a controversial report, Paradise and Bess (1994) disagreed with the recommendation for implementation of universal newborn hearing screening programs. The authors posed questions regarding the sensitivity and specificity of currently available technology. They were concerned that there might be an unacceptably high number of over-referrals for follow-up testing. The authors also questioned the availability of qualified professionals and facilities to provide follow-up diagnostic testing that a universal screening program would generate. In addition, they did not feel confident that enough qualified professionals were available to provide appropriate treatment for all infants identified with hearing loss.

Although many of the opinions of Paradise and Bess have been addressed in the literature, the issue of the availability of facilities and qualified professionals has not been put to rest. Yoshinaga-Itano (1999) questioned the availability of audiologists with expertise in pediatric evaluation and treatment. Northern and Hayes (1994) and White and Maxon (1995) felt that there may

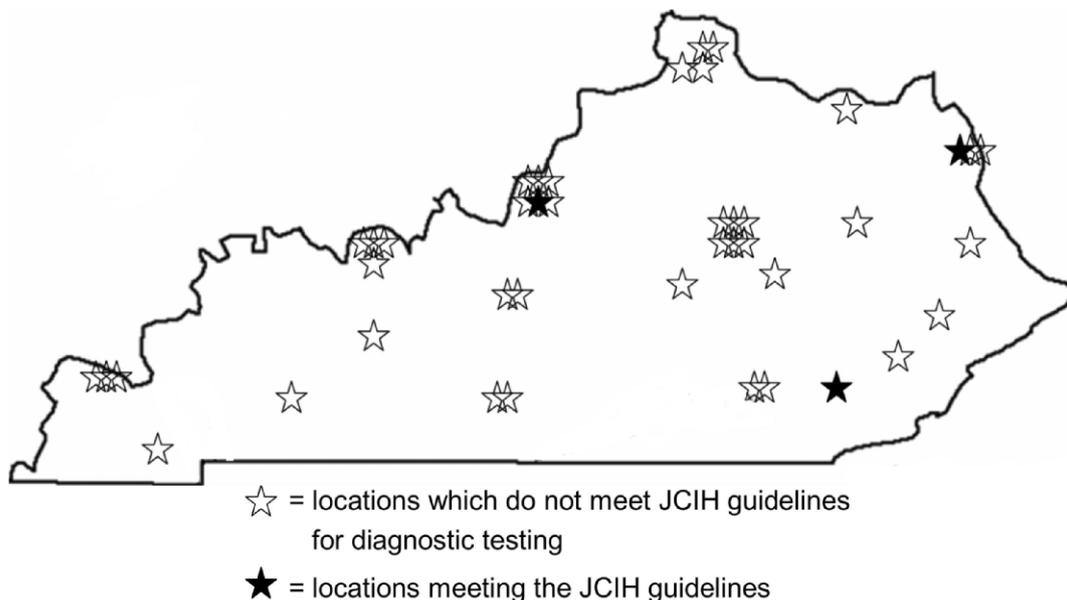
be issues with the availability of qualified facilities and professionals available to provide follow-up services, but this should not be reason to delay the establishment of universal newborn hearing screening programs. Nonetheless, without adequate follow-up services, the benefits of UNHS are lost. Therefore, the next step in assuring success in newborn screening programs is the availability of appropriate facilities and professionals to provide diagnostic testing in a manner consistent with the Joint Committee guidelines.

### DIAGNOSTIC FOLLOW-UP TESTING IN KENTUCKY

In July 2000, the Commonwealth of Kentucky enacted a law requiring that all hospitals in the state with more than 40 births per year perform a hearing screening on every newborn. The purpose for this legislation was to facilitate the identification of hearing loss at birth. Kentucky's program is titled "Kentucky Infant's Sound Start" (KISS) and is managed by the Commission for Children with Special Health Care Needs (CCSHCN). The legislation also calls for the establishment of an advisory committee,

referral for comprehensive audiologic evaluation with a failed screening, and requirements for management of data associated with the program. Kentucky's screening program has been in place for four years and, reportedly, screened 99% of the infants born in the state in 2003 (National Center for Hearing Assessment and Management [NCHAM] Web site: [www.infanthearing.org](http://www.infanthearing.org)). Currently, Kentucky is one of 37 states to have passed legislation requiring universal newborn hearing screening.

The purpose of hearing screening at birth is to assure early identification of hearing loss. The goal of identifying hearing loss by three months of age requires that infants failing the hearing screening be referred to centers that are capable of providing the necessary diagnostic services. Though it has been reported that 99% of all infants born in Kentucky are screened at birth, no information is readily available regarding the accessibility and provision of the diagnostic testing for those infants failing the screening. The KISS Web site (<http://chs.state.ky.us/commissionkids/unhs.htm>) lists 42 centers distributed across the commonwealth available to perform the diagnostic testing (Figure 1). This list of sites



**Figure 1.** Locations of the centers listed on the Kentucky Infant's Sound Start (KISS) Web site as providing follow-up diagnostic audiology services.

includes the address and phone number but no other information regarding the technology, expertise, or adherence to recommended guidelines is provided. Therefore the purpose of this project was to determine the extent to which the Commonwealth of Kentucky is meeting the JCIH guidelines for diagnostic testing on infants failing hearing screening at birth. Specifically, the purpose of this project was to survey those centers that are listed as the providers of diagnostic services to determine the extent to which various technologies are employed during the diagnostic process, the availability of intervention programs at these centers, and the relative expertise of the staff in providing both diagnostic and intervention services.

## METHODS

A survey tool was developed to assess the procedures and protocols utilized by facilities listed on the KISS Web site in providing diagnostic testing and subsequent referrals and intervention services for newborns failing the required hearing screening. The survey was based on the diagnostic and intervention guidelines from the JCIH *Early Hearing Detection and Intervention* document. Forty-two facilities listed on the KISS Web site as a “Kentucky Diagnostic Audiology Resource Center” were contacted by telephone in an attempt to complete the survey.

The survey included questions regarding equipment and procedures used for the audiologic evaluation of the newborn. Specific questions were developed to survey use of history taking, acoustic immittance measures, otoacoustic emissions, auditory brainstem response, frequency-specific evoked potentials, bone-conducted ABR, auditory steady state response (ASSR) audiometry, and behavioral audiometry. Questions related to test protocols for each of the specific tests as well as the availability of equipment were included.

Questions regarding referrals following identification of hearing loss were also included. Referrals to the primary care physician, to the otolaryngologist, and for genetic evaluation are recommended by the JCIH. The survey also sought to determine the extent to which the KISS report form was completed and returned to the CSHCN

UNHS data management program for infants receiving audiologic evaluation.

The survey questions related to intervention included information regarding the provision of amplification for infants and children and follow-up services. The availability of cochlear implant services was also surveyed. Due to the JCIH’s recommendation for referral of infants identified with hearing loss to a public agency, survey also asked questions regarding the Kentucky’s Early Intervention Program (First Steps). The survey was conducted in the spring of 2004.

## RESULTS

Contact was made with 40 of the 42 facilities listed on the KISS Web site as Kentucky Diagnostic Audiology Resource Centers. Of the two sites that were not contacted, one site was no longer in business, and one could not be reached to complete the survey. Of the 40 centers contacted, one facility reported having no audiology services available, and two facilities reported having no current on-site audiology services. Three of the 40 facilities reported that follow-up evaluations for infants referred from the UNHS programs were not conducted at their sites. The remaining 34 facilities completed the survey. In summary, seven centers listed on the KISS Web site no longer conducted follow-up evaluations of newborns failing infant screenings, and one was unavailable to complete the survey. The 34 centers completing the survey represent 81% of the centers listed on the KISS Web site but 95% of the centers listed that provide follow-up services.

Each center was asked to estimate the total number of diagnostic hearing evaluations performed at their facility that were referred from UNHS failures during the last calendar year. One site reported no evaluations were performed over the past year. Seventeen of the sites reported performing fewer than 25 evaluations per year. Three sites reported 11–25 evaluations, and six sites reported doing 26–50 evaluations per year. There were five facilities that reported doing 51–100 evaluations, and three sites that estimated doing more than 100 evaluations during the last year. Two facilities indicated that they were unable to estimate

the number of evaluations that were performed during that calendar year.

The first group of questions in the survey related to the components of the diagnostic protocol used for audiologic evaluation following referral from a failed UNHS. Table 1 lists the questions and cumulative responses from the 34 facilities. A child and family history was obtained by all 34 (100%) of the participating facilities. Tympanometry is routinely conducted at 31 (91%) of the surveyed facilities. Of those facilities that routinely use tympanometry, only 12 reported routinely using a high-frequency probe tone for infants under six months of age. This represents about one-third (35%) of the 34 facilities. Acoustic reflexes were routinely tested at 16 (47%) of the facilities. Of the 16 sites that obtain acoustic reflexes, five use only screening techniques rather than obtaining acoustic reflex thresholds, and seven of the facilities obtain thresholds, while four sites reportedly use both techniques. At the most, only 11 (32%) of the 34 facilities contacted obtain acoustic reflex thresholds as part of the diagnostic protocol.

Conversely, otoacoustic emission testing is conducted at 32 of the 34 of the surveyed sites (94%), with 24 facilities using distortion product otoacoustic emissions (DPOAEs) and

three using transient evoked otoacoustic emissions (TEOAEs). There were five sites that had both DPOAEs and TEOAEs. The two sites that do not use OAE testing reported not having OAE equipment available. Those reporting use of DPOAEs were asked about the intensity levels routinely used for testing. The majority (23 out of 34) reported using 65/55 dB as the intensities of the stimulus tones, while six others were unsure of the levels used for stimulation. Five of the facilities using the 65/55 protocol reported that if responses were not obtained to these stimulus levels, a 70/70 protocol is then used.

The final survey questions related to components of the standard diagnostic protocol were the facilities' use of electrophysiologic measures, such as the auditory brainstem response (Table 2). Of the 34 respondents, 24 (71%) reported using ABR procedures as a part of the follow-up evaluation, while ten centers do not routinely use the ABR procedure. Eight of the ten sites do not have ABR equipment, and two facilities do not perform threshold searches, using their equipment only for adult retrocochlear diagnostics. Five of the 24 sites perform ABR measures only if appropriate responses to OAE testing are not obtained. Therefore, 19 of the 34 sites (59%) surveyed routinely

**Table 1. Are the Following Tests Included in Your Facility's Protocol for Audiologic Evaluation following a Failed Newborn Hearing Screening?**

DIAGNOSTIC TEST	YES	NO
Child and family history	34 (100%)	0 (0%)
Tympanometry (226 probe tone)	31 (91%)	3 (9%)
Tympanometry (high-frequency probe tone)	12 (35%)	22 (65%)
Acoustic reflex thresholds	16 (47%)	18 (53%)
Otoacoustic emissions	32 (94%)	2 (6%)
ABR (Electrophysiologic tests)	24 (71%)*	10 (29%)
Behavioral audiometry	29 (85%)	5 (15%)

Note: N = 24.

\*Of the 24 sites reporting use of ABR, five sites use ABR only if responses to OAEs could not be obtained.

**Table 2. Responses to Questions Regarding Electrophysiologic Test Measures Used in the Evaluation of Newborns**

ELECTROPHYSIOLOGIC PROCEDURE	YES	NO
Are ABR click thresholds obtained?	24 (71%)	10 (29%)
Are frequency specific ABR stimuli routinely used?	10 (29%)	24 (71%)
Polarity change to assess for auditory neuropathy?	10 (29%)	24 (71%)
Is bone conduction routinely used?	10 (29%)	24 (71%)
Do you have ASSR available?	3 (9%)	31 (91%)
Do you use ASSR in the evaluation of newborns?	0 (0%)	34 (100%)

Note: N = 34.

conduct ABR evaluations as part of the diagnostic protocol.

All centers performing the ABR procedures (24) report obtaining thresholds for click stimuli. Clicks have long been used to measure hearing sensitivity and to make decisions regarding hearing loss for high-frequency sounds. Thresholds, the lowest intensity level that evokes an electrophysiologic response, are established to judge the presence versus absence of hearing loss. Interestingly, the intensity level used to benchmark normal auditory function varied among the facilities. One site considered a response at 40 dB or less as normal; nine considered 30 dB as normal; five centers called 25 dB normal; seven called 20 dB normal; and two sites considered 15 dB to be normal.

Over the past decade, auditory neuropathy has come to be recognized as a significant auditory system disorder. This disorder is a complex condition characterized by a unique constellation of auditory test results. Auditory neuropathy requires nontraditional intervention methods, and therefore differentiation of the condition is critical to management. The differential diagnosis is based on simple manipulations of the click stimuli for the ABR. The manipulations include inverting the polarity of the stimuli to determine if significant changes occur in the electrophysiologic response. Ten (42%) of the facilities using ABR routinely use high-intensity stimuli of inverted polarities to search for the specific pathology of auditory dys-synchrony. Fifty-eight percent (14) of the facilities report not incorporating varied polarities as part of their protocol.

The Joint Committee recommends the use of frequency specific stimuli to determine hearing losses not apparent through use of clicks and to define the configuration of hearing loss for individuals identified with hearing loss. Frequency specific stimuli (i.e., tone bursts, tone pips) are routinely performed at only ten (42%) of those facilities that perform ABR click thresholds. This represents less than one in three of the 34 sites contacted. Six of the 14 sites (43%) stated that they do not conduct frequency-specific thresholds, reportedly as they do not have the equipment available to conduct such a test. The remaining eight sites (57%) have the capability of performing frequency-specific

tests but do not use it as part of the diagnostic evaluation on infants failing the newborn screen. Among the reasons expressed as to why frequency-specific stimuli are not routinely utilized were not feeling comfortable conducting the test and/or interpreting the results, and not having the time to complete the test while the baby is sleeping.

Bone-conducted ABR thresholds, used to identify hearing losses associated with middle or external ear disorders, are routinely obtained by only 10 of the 34 sites (29%). Seven of the sites that do not perform this test do not have the equipment necessary to do bone-conduction ABR.

Auditory Steady State Response (ASSR) is a relatively new electrophysiologic procedure that is being integrated into the diagnostic test battery to determine the presence of hearing loss. ASSR provides frequency specific information that would meet the Joint Committee guidelines. Three of the 34 facilities surveyed in Kentucky have the ASSR equipment (Table 2); however, there are no facilities that are routinely including this test in the follow-up evaluation of infants referred for the UNHS program at this time.

The varied procedures used to test hearing in newborns is facilitated by having the infant sleep. Testing is either unduly prolonged or delayed for the nonsleeping infant. The use of sedation for testing was surveyed, and 9 of the 24 facilities that conduct electrophysiologic measures use sedation to facilitate the diagnostic testing. This represents approximately 38% of all facilities in Kentucky conducting follow-up testing.

Twenty-nine facilities reported behavioral testing as part of the standard protocol. However, the next question addressed the age at which behavioral testing was first conducted. No consistent pattern of responses was obtained although 14 centers reported initiating behavioral testing at six months of age, and 15 centers reported beginning behavioral testing between birth and six months of age. The rest reported various ages ranging from nine months to three years of age.

The survey attempted to determine how the results of the diagnostic evaluation were reported. Though not required by law, the reporting of results to the KISS program is encouraged. Twenty-three of the 34 facilities reportedly send results to the KISS data

management resource (Table 3). Eighty-eight percent (30 of 34 facilities) routinely contact the child's primary care provider with the results of the evaluation. Twenty-seven (79%) of the facilities routinely refer a child identified with hearing loss to an otolaryngologist. In addition, 5 of the 34 facilities are part of an otolaryngology practice and therefore involve the otolaryngologist in the care of the child. Therefore, 32 of the 34 facilities routinely involve an otolaryngologist in the care of infants identified with hearing loss. However, only 14 of the 34 facilities (41%) routinely refer infants identified with hearing loss for a genetic evaluation.

The final section of the survey inquired about the provision of intervention services for infants identified with hearing loss. Twenty-three of the 34 facilities (68%) provide intervention services. However, only 19 of these 23 facilities dispense amplification devices for children identified with hearing loss. Facilities were then asked to identify the percentage of infants who, when identified with hearing loss prior to six months of age, are then fit with hearing aids by six months of age. Six facilities indicated that 100% of the children were fit by six months; three facilities indicated that 80–90% were fit by six months; two facilities reported fitting 50% of the children; and three indicated that no child was fit by six months of age. Four centers indicated they had not identified a child with hearing loss at the time of the survey. Regarding the provision of cochlear implant services, only 4 of the 34 centers (12%) reported providing cochlear implant services, but 26 of the 34 facilities indicated they refer children for cochlear implant services to other centers.

## DISCUSSION

It is well accepted that children identified with hearing loss who receive intervention prior to six months of age have significantly better outcomes than those who are later identified (Yoshinaga-Itano et al, 1998). Important for early intervention is the early identification of infants with hearing loss. Thus, guidelines for the implementation of a hearing screening program for newborns are provided in the Joint Committee on Infant Hearing document. Recommendations for program development, the personnel required for a successful program, the technologies to be employed, as well as the screening protocols are included in the JCIH guidelines. Further, benchmarks and quality indicators for the screenings are provided. These include numbers of infants to be screened, ages of the infants to be screened, recommended referral rates and documentation of the infants screened and tracking information on those who are referred for further evaluation.

Beyond the screening component, the JCIH document provides guidance for the provision of the follow-up diagnostic testing, including the scope and range of procedures that should be included in the diagnostic protocol. The JCIH recommends a test battery that includes physiologic measures as well as developmentally appropriate behavioral tests. The purpose of this battery is to assess the integrity of the auditory system, obtain ear-specific estimates of type, degree, and configuration of hearing loss, and develop appropriate intervention options.

In Kentucky, universal newborn hearing screening has been conducted since July 2000. This program has been developed and implemented in virtually all birthing hospitals in the state. As previously noted, it

**Table 3. Responses to Questions Regarding Typical Referrals following Failure of Newborn Hearing Screening and Subsequent Identification of Hearing Loss**

REFERRAL	YES	NO
Primary care physician	30 (88%)	4 (12%)
Otolaryngologist	32 (94%)	2 (6%)
Genetic evaluation	14 (41%)	20 (59%)
KISS form	23 (70%)	11 (30%)

Note: N = 34.

has been reported that 99% of all infants born in the state are screened for hearing loss. Early identification of hearing loss, however, is not limited to the development and implementation of a successful screening program. The screening program itself only identifies those newborns in need of diagnostic testing. It is the diagnostic stage of the process that actually identifies newborns with hearing loss. Therefore, the diagnostic component is a fundamentally important component to successful outcomes of children with congenital or early onset hearing loss.

The purpose of this study was to evaluate the degree to which facilities in Kentucky that provide the important follow-up diagnostic services are meeting the JCIH guidelines and protocols. The KISS Web site lists 42 facilities in the commonwealth that provide the audiologic diagnostic services. Forty of the 42 sites were contacted with a survey to determine the protocols used for the evaluations. Although the KISS Web site lists 42 facilities, only 34 currently provide audiologic diagnostic services. It is likely that there are other facilities in Kentucky that are providing follow-up diagnostic services but are not listed on the KISS Web site.

All facilities in Kentucky reported conducting a case history as part of the diagnostic protocol. Most facilities also reported conducting tympanometry. The use of a high-frequency probe tone is advantageous in conducting tympanometry in infants under six months of age (Sininger, 2003), but only about one-third of the centers in Kentucky used this procedure. Acoustic reflexes were part of the diagnostic protocol in less than half of the centers, and in less than one-third were thresholds actually established. The other centers screened the acoustic reflexes.

Otoacoustic emissions are routinely conducted in most centers as part of the evaluation of infants failing a newborn screening. The only centers not conducting OAEs did not have the equipment available. The majority use DPOAEs with the most common stimulation intensities for F1 and F2 being 65 dB and 55 dB respectively. It is interesting to note, however, that a substantial number of facilities were unsure of the intensity settings.

Nineteen of the 33 sites surveyed routinely conduct electrophysiologic measures

as a part of the follow-up evaluation. This represents slightly more than half (58%) of the designated sites in Kentucky. All of these sites reported obtaining click thresholds as a part of the test battery. However, the threshold level that is considered to represent normal hearing varies widely among the sites. Results of this survey indicated that only two sites specify 15 dB or below as normal. Prior studies have shown that a significant number of infants with hearing loss will be missed when using click thresholds of 20 dB and above as indicative of normal hearing. Interestingly, more than half the sites in Kentucky conducting click-evoked ABR report specifying responses between 25 dB and 40 dB as normal.

Additional electrophysiologic measures, including assessment of the integrity of the auditory system through inversion of polarity to search for auditory dys-synchrony, use of frequency specific tonebursts, and bone-conducted ABR thresholds are recommended by the JCIH as necessary components of the test battery. Only 10 of the 23 sites that perform ABR testing include these additional tests. Therefore, less than 25% of the sites listed as providing evaluation following referral from UNHS are performing the recommended electrophysiologic test procedures.

There were no sites using the ASSR technology; however, this is not surprising due to the recent availability of equipment for use in clinical settings. In addition, the Joint Committee guidelines were issued prior to the availability of ASSR. The use of this procedure, however, would allow a site to conduct the necessary frequency-specific electrophysiologic test procedures to meet the Joint Committee guidelines. Regardless, the availability of tone-burst stimuli using conventional ABR procedures does allow clinics to meet the frequency specificity guideline.

The JCIH statement refers to the use of "observation of the infant's behavioral response to sound" as a part of the test protocol for follow-up evaluations. This statement could be interpreted as performing behavioral audiometry (e.g., behavioral observation audiometry, visual response audiometry, conditioned oriented response) as a part of each evaluation. However, it may also be interpreted as an informal observation to the infant's response to sound. The question

was initially included in the survey as referring to formal audiometry, but the responses suggested that the audiologists did not consider the behavioral component as part of the test battery for evaluations but, rather, as a stand-alone entity that can be used in place of electrophysiologic measures. The majority of the sites that use ABR measures as a component of the test battery do not include formal audiometry as a part of the evaluation. Most of the sites that do not have ABR but do conduct diagnostic evaluations are using formal audiometry as a part of the test battery. This is concerning because behavioral audiometry conducted with infants under five months developmental age is typically not reliable and does not provide the necessary information related to type, degree, and configuration of hearing loss. Gravel and Hood (1999) contend that reliable behavioral testing can first be performed at the developmental age of 5–6 months. In Kentucky, 15 sites report using formalized behavioral testing on infants less than six months of age.

In summary, the JCIH position statement recommends that the test battery used for audiological evaluation following referral from the UNHS program be used to assess the integrity of the auditory system, assess hearing sensitivity, and identify intervention options. To accomplish this, the test battery must include child and family history, electrophysiologic measure of threshold, frequency-specific stimuli, bone-conducted threshold, a measure to detect auditory neuropathy, a measure of middle ear function using the appropriate frequency probe stimuli, acoustic reflex thresholds, OAEs, and observation of the infant's response to sound. There are 42 facilities in Kentucky listed on the KISS Web site as centers that conduct audiological evaluation following referral from UNHS. When taken in total, only 3 of the 42 sites appear to provide the full range of tests and procedures recommended by the JCIH. Of interest, however, is that two of these three sites report providing more than 100 evaluations per year, which suggests the proportion of infants receiving follow-up testing that meets the Joint Committee guidelines is actually higher than would otherwise be surmised. Conversely, one of the sites that provides more than 100 evaluations per year does not meet the Joint Committee guidelines.

The JCIH recommendations also include referral by the diagnostic audiologist to the "medical home," or primary care physician, and to an otolaryngologist. Table 3 shows the results of the responses to the questions regarding referral of infants identified with hearing loss. Eighty-eight percent of the sites report contacting the infant's "medical home," or primary care physician, either by phone contact or sending a report containing the results of the evaluation. The majority of the sites (94%) typically either refers to an otolaryngologist or is employed by an otolaryngologist and performs the tests in the otolaryngology office. The JCIH document recommends referral for genetic evaluation when necessary; however, it indicates that the otolaryngologist will make that recommendation. The survey indicated that 42% of the audiology facilities providing diagnostic evaluations also provide genetic referrals when necessary. Seventy-six percent of the facilities providing audiological evaluations report that they will refer infants who are possible candidates for cochlear implantation to the appropriate facilities that provide those services.

Of particular interest is the rate at which the results are reported to the KISS program. Only 23 of the 34 sites (70%) send results to the program. Therefore, data from nearly one-third of all diagnostic facilities tested in follow-up may not be available in a centralized database.

Of the 34 facilities surveyed, 23 sites report that they provide early intervention services. Nineteen of those sites report that they dispense amplification to infants referred from the UNHS program. A significant number of those sites indicated that though they are willing to dispense to very young infants, it is very infrequent that they have the opportunity to do so. According to the JCIH document, infants with hearing loss should be fit with appropriate amplification before the age of six months in order to provide for the best outcome. There were six sites that reported consistently fitting infants before the age of six months, if they are identified by that age. However, these same sites also reported that it is not often that the necessary information for the fitting of amplification is obtained before the age of six months. Many sites referred to the large amount of administrative work that must be completed before procuring the hearing

aids, which often delays the fittings to significantly greater than six months of age as well. This too causes concern in regards to the follow-up of infants referred from the UNHS. Without appropriate diagnostic results by the age of three months as recommended by the JCIH, appropriate intervention is not likely to be provided by the age of six months.

## CONCLUSIONS

Presuming that the JCIH guidelines reflect the appropriate protocols and procedures to provide diagnostic audiologic services to infants who are referred from the UNHS program, the results of this study demonstrate that only 3 of 42 centers in Kentucky provide appropriate levels of service. It appears that Kentucky's UNHS program is accomplishing the goal of screening infants and making referrals for evaluation by one month of age; however, it seems that without access to appropriate diagnostic services, the benchmark of identification by the age of three months cannot be achieved. While the age of identification has most certainly been reduced in recent years, the goal is to meet the guideline of identification by three months of age. If infants with hearing loss cannot be identified by three months of age, it is also likely that the goal of intervention by six months of age is also not being met. Therefore, in spite of a successful UNHS program, this study suggests that adequate services beyond the screening component are not available and thus compromising the potential success of children with congenital and early onset hearing loss.

Thus, the conclusions of Bess and Paradise regarding the availability of technologies, facilities, and professionals to conduct the follow-up testing on infants failing a newborn screening program continues to be accurate, at least in Kentucky. It is entirely likely that Kentucky is representative of many states, and therefore the availability of follow-up services in other states is likely open to question. Certainly, the lack of access to appropriate diagnostic testing reduces the value of the UNHS. It would obviously be interesting to evaluate the status of the availability and utilization of follow-up services in other states.

Over the past decade, much time and energy has been devoted to the development and implementation of a UNHS programs across the country. Assuming Kentucky is representative of the availability of diagnostic follow-up services, it is now time for hearing health-care professionals to devote time and energy to the development of the next stage in the process and overcome the inadequacies found in this study. These professionals include, but are not limited to, audiologists, primary care physicians, otolaryngologists, nurses, and other medical professionals who are providing services to infants.

These services might be improved in a number of ways. First, facilities must be identified that desire to and are able to provide the full scope of audiologic diagnostic services. The number of sites providing evaluations could be consolidated in order to have consistent and complete diagnostic services provided by experienced clinicians. Secondly, clinicians providing the diagnostic services should be trained in the latest procedures and protocols. Using the JCIH guidelines, training should include the test battery protocols and how to perform each test and interpret the results. Over the past decade, training has focused on the provision of screening services rather than the follow-up services. In spite of the Bess and Paradise cautions, there may be a more widespread presumption that there is less need for training in follow-up audiologic testing. Finally, the equipment necessary for appropriate testing must be available at the facilities that are providing the follow-up evaluations.

Once the identification benchmark of three months of age is met, intervention by six months of age will be attainable. As mentioned previously, fitting of pediatric amplification requires expertise very different from the fitting of adult amplification. Therefore, consolidation of sites, training of audiologists who are fitting infants, and appropriate equipment for the fitting of infants should be provided.

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