Newborn Hearing Screening Speeds Diagnosis and Access to Intervention by 20–25 Months

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Abstract

Background: Newborn Hearing Screening (NHS) programs aim to reduce the age of identification and intervention of infants with hearing loss. It is generally accepted that NHS programs achieve that outcome, but few studies have compared children who were screened to those not screened in the same study and during the same time period. This study takes advantage of the emerging screening programs in California to compare children based on screening status on age at intervention milestones.

Purpose: The purpose of this study was to compare the outcomes of cohorts of children with hearing loss, some screened for hearing loss at birth and others not screened. Specifically, the measures compared are the benchmarks suggested by the Joint Committee on Infant hearing for determining the quality of screening programs.

Study Sample: Records from 64 children with bilateral permanent hearing loss who were enrolled in a study of communication outcomes served as data for this study. Of these children, 47 were screened with 39 failing and 8 passing, and 17 were not screened.

Intervention: This study was observational and involved no planned intervention.

Data Collection and Analysis: Outcome benchmarks included age at diagnosis of hearing loss, age at fitting of amplification, and age at enrollment in early intervention. Delays between diagnosis and fitting or enrollment were also calculated. Hearing screening status of the children included screened with fail outcome, screened with pass outcome, and not screened. Analysis included simple descriptive statistics, and t-tests were used to compare outcomes by groups: screened/not screened, screened pass/screened fail, and passed/not screened.

Results: Children with hearing loss who had been screened as newborns were diagnosed with hearing loss 24.62 months earlier, fitted with hearing aids 23.51 months earlier, and enrolled in early intervention 19.98 months earlier than those infants who were not screened. Screening status did not influence delays in fitting of amplification or enrollment in intervention following diagnosis. Eight of the infants with hearing loss (12.5%) passed the NHS, and the ages at benchmarks of those children were slightly but not significantly earlier than infants who had not been screened.

Conclusions: The age at achievement of benchmarks such as diagnosis, fitting of amplification, and enrollment in early intervention in children who were screened for hearing loss is on target with stated goals provided by the Academy of Pediatrics and the Joint Committee on Infant Hearing. In addition, children who are not screened for hearing loss continue to show dramatic delays in achievement of benchmarks by as much as 24 months. Evaluating achievement of benchmarks during the start-up period of NHS programs allowed a direct evaluation of ability of these screening programs to meet stated goals. This demonstrates, unequivocally, that the NHS process itself is responsible for improvements in age at diagnosis, hearing aid fitting, and enrollment in intervention.

Key Words: Early intervention, newborn hearing screening

Abbreviations: ABR = auditory brainstem response; JCIH = Joint Committee on Infant Hearing; NHS = newborn hearing screening; NICU = neonatal intensive care unit; OAE = otoacoustic emission; UNHS = universal newborn hearing screening

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Universal newborn hearing screening (UNHS) is now implemented throughout the United States and around the world and is generally accepted by pediatricians and primary care physicians (Moeller et al, 2006). The goal of newborn hearing screening (NHS) is earlier identification of hearing loss, which leads to earlier intervention and, in turn, facilitates the development of communication and auditory skills in children with hearing loss.

The most important outcome of early identification would be improved communication ability, including language as well as auditory-based outcomes such as speech perception and speech production. Studies supporting the relationship between early detection and improved communication ability are still emerging. The relationship between early identification/intervention and language is most often studied, and such studies have consistently shown that earlier intervention provides a significant advantage in language development for children with hearing loss (Robinshaw 1995; Yoshinaga-Itano et al, 1998; Moeller 2000; Kennedy et al, 2006).

In addition to communication outcomes, however, it is important to document that screening for hearing loss actually decreases the age at which important benchmarks are achieved for children with hearing loss. The Joint Committee on Infant Hearing (JCIH) Year 2000 Position Statement listed benchmark ages for early hearing detection and intervention systems to judge program quality. These benchmarks are (1) beginning audiologic evaluation before three months of age, (2) use of amplification within one month of confirmation of hearing loss, and (3) enrollment in early intervention programs before six months of age. These benchmarks insure that follow-up of the early suspicion of hearing loss, from failed newborn screening, is accomplished in a timely manner. Without achieving such goals, the more long-term benefits of early identification cannot be realized.

**PRIOR TO UNHS**

Studies conducted before the advent of widespread UNHS evaluated benchmarks similar to those suggested by JCIH. These studies generally found that factors such as high-risk indicators and degree of hearing loss influenced the outcomes. For example, Mace et al (1991) evaluated medical records of 123 children followed for hearing loss during a time period prior to UNHS. Although identification was found to be earlier for children with greater severity of hearing loss, the overall median age of identification (diagnosis) was 2.1 years. For children with unilateral or borderline (15–25 dB HL) hearing loss, the age of identification was 4.0–5.7 years of age. This study also found that risk factors such as known syndromes or craniofacial anomalies and other handicapping conditions led to earlier diagnosis in general, but otitis media did not influence age of identification.

A few years later, Harrison and Roush (1996) analyzed survey data from parents of children with hearing loss who were enrolled in early intervention programs, also during a time period prior to UNHS in most states. They determined that hearing loss was diagnosed in children without risk factors at approximately 22 months of age if the loss was mild to moderate but at 13 months of age if the loss was severe to profound. Intervention and hearing aid fitting were affected in similar ways with each happening at about 28 months of age for children with milder losses and 16 months for those with severe to profound losses. For children with known risk factors for hearing loss, the effect of degree of loss on identification age was not seen. The children with risk factors were identified at about 12 months of age, but fitting of amplification was earlier for those at-risk children with severe to profound loss (15 months) than those with mild to moderate (22 months) hearing loss.

**STUDIES OF UNHS**

The Wessex Study (1998), conducted in England, compared benchmarks for infants born in hospitals that screened for hearing loss using neonatal otoacoustic emissions (OAEs) and auditory brainstem response (ABR) to those born in hospitals without neonatal screening procedures but screened at home by conventional (distraction) tests. Each group was evaluated simultaneously during a period of time prior to widespread UNHS in England. Results from this study indicated that a greater proportion of the children exposed to neonatal screening were confirmed and intervention (management) was begun before 10 months of age when compared to children born in hospitals without screening. This study does not allow for direct comparison of ages when specific benchmarks were reached; rather, it compares the two groups to determine whether NHS had significant effect on the percentages of infants reaching some of the benchmarks.

In an evaluation of UNHS in New York State, Dalzell et al (2000) found the median age of diagnosis to be three months for a group of 85 infants identified by NHS. Thirty-six of the infants had been fitted with amplification at a median age of 7.5 or 4.5 months following diagnosis. The remaining infants were not necessarily delayed; rather, they were not fitted with amplification due to factors such as unilateral hearing loss, mild loss, or illness. This study found that more than half of the infants identified with hearing loss were enrolled in intervention by five months of age, and the median age of intervention was three months. It is noted, however, that some of those infants were enrolled in early intervention before tests to confirm diagnosis were completed.

Uus and Bamford (2006) also conducted a survey of 169 infants identified by NHS with bilateral hearing loss in Britain. This study found that overall the median age at diagnosis was ten weeks. Diagnosis age
was slightly greater for the neonatal intensive care unit (NICU) population. The median age at enrollment of early support was also found to be 10 weeks, and the median age at fitting of hearing aids was 16 weeks, also slightly later for infants in the NICU. The median delay of fitting was five weeks.

Clearly, the median age at which infants achieve important benchmarks including firm diagnosis of degree and type of loss, fitting of amplification, and enrollment in early intervention seems to decrease, from about two years to about three months or less when infants are identified through an NHS program. The New York and English studies identified and followed subjects who had failed NHS only. The question remains whether the passage of time has increased the awareness and zeal of those involved in early detection and intervention for hearing loss and, if so, whether the reported decreases in age of benchmark achievement is due to the screening process itself. The study described here takes a different approach that allows a direct comparison of benchmarks achieved for children who were screened for neonatal hearing loss and those who were not. Subject selection for this study started with infants and children at the time of audiologic diagnosis of bilateral, permanent hearing loss regardless of NHS status or age at diagnosis. These children and their families were recruited for a longitudinal study of auditory-based communication outcomes in infants and toddlers with hearing loss (Sininger et al, 2007). Children identified with hearing loss before the age of 5 years are being followed to evaluate the development of language (primarily spoken language), speech production, and speech perception. Information regarding the status of NHS on each child was obtained during the enrollment into the study.

This project identified a cohort of infants and toddlers with hearing loss who displayed a variety of histories related to NHS. Some were screened and some not screened. Of those screened in the newborn period, some passed the screening while others failed. This study evaluates children’s outcomes on JCIH and other benchmarks as related to screening status. This approach allows evaluation of those infants who initially passed screening but later were found to have hearing loss. This is not possible in a study that follows only the failed screenings. This approach also allows for a control group of subjects that were not screened.

Subjects for this study were enrolled between 2002 and 2005 to take advantage of the staggered start-up of NHS in California. In 1998, legislation was enacted in California requiring screening of newborns for hearing loss but only in those hospitals with licensed perinatal units that were participating in the California Children’s Service (CCS) program. This distinction described the birthing hospitals for approximately 70% of infants born in California. The legislation called for hospitals to be certified for participation in the program starting in July of 1999 but no later than January 1, 2003. Not all hospitals were screening infants for hearing loss when the study began, and the selection of screened and not-screened subjects was random. This is important given the ethical issues that would prevent a random assignment to such groups in a prospective study.

**METHOD**

**Subjects**

A total of 63 children have been enrolled in the longitudinal study. The children were born between 1996 and 2004, and their families were recruited to the study between 2002 and 2005. Each subject was recruited with the aid of clinical audiologists no more than six months following diagnosis of hearing loss and generally at the time of fitting with amplification. Children and their families were enrolled with cooperation of audiology diagnostic clinics located at UCLA Medical Center and the House Ear Institute CARE Center, both in Los Angeles, or at the Providence Hearing and Speech Center in Orange, California. Recruitment and informed consent of UCLA and Providence patients/families was overseen by the UCLA Institutional Review Board and House patients/families by the St. Vincent’s Medical Center Institutional Review Board.

NHS was not a prerequisite for enrollment in the study, and screening was not available at all birthing hospitals in California. All children identified with bilateral sensorineural hearing loss requiring amplification were offered enrollment in the study. The timing of enrollment of subjects for this study was designed to take advantage of the start-up of NHS in California assuming that subjects would be selected from a wide range of ages of diagnosis. The primary hypothesis of the longitudinal study is that the age at which amplification is fitted will be an important predictor of auditory-based outcomes. The advantage of sampling from a population in which screening was not fully implemented is that there is a range of screening experiences and, consequently, a range of ages at intervention including fitting of amplification. Such a distribution of ages facilitates hypothesis testing.

The criterion for enrollment in the study was a diagnosis of bilateral permanent hearing loss sufficient to warrant fitting of amplification. Exclusion criteria included any significant motor, vision, or developmental involvement sufficient to influence development of spoken language. In some cases, such disorders were discovered after enrollment, and these subjects were dropped from the overall study. However, the outcomes described in this manuscript include all subjects for whom data are available regardless of whether the subject continued in the longitudinal study. Children with auditory neuropathy were also excluded from the study, and none were enrolled.

The subject pool for this study included 33 males and 30 females, with a mean gestational age of 38.14 weeks (s.d. = 2.47), birthweight, of 3117.6 grams (s.d. = 765 grams), and
socioeconomic status, as determined by mother’s years of education, of 15 years (s.d. = 3.22). The degree of hearing loss in the better ear ranged from mild to profound.

Data Collection

All data reported were gathered from questionnaires, enrollment forms, and audiologic chart records. Parents reports were verified as necessary. NHS in California is closely regulated by the state. Participating hospitals are required to report all screening failures to the appropriate hearing coordination center (HCC) and to make immediate referral to an approved audiology center. The audiologic records of infants who failed NHS include state forms for reporting outcomes of the audiologic testing to the California HCCs. However, this process aided in documenting screening failures only, as passed infants are not reported by name. If parents reported a screening pass, that result could be verified through the HCC, which would have documentation of the birth hospital’s screening status at the time of the infant’s birth, and if a fail was not reported for that child, a pass was presumed. The HCC would also have records of missed infants. If parents reported that their infant was not screened, the screening status of the birth hospital at the time of the birth could be verified through the HCC. Dates of important benchmarks were recorded and converted to ages for analysis. Data gathered included:

1. **Screening Status** including screened with pass outcome, screened with failed outcome or not screened. The outcome is that recorded at discharge from the birth admission hospital. According to California regulation, any infant who fails in one or both ears receives a second screening before discharge. Both ears are rescreened regardless of initial outcome. If both ears pass on screen two, a pass is recorded, otherwise a “fail” or “refer” is recorded. The distinction between a bilateral and unilateral fail was not recorded for this study. The technology used for the screening procedure was not noted in the infants’ records and generally was not recalled by parents. By California regulation, hospitals may choose to screen with auditory brainstem response (ABR) or distortion product or transient otoacoustic emissions (OAEs) administered by an appropriate infant screening device. Screening information was gleaned from parent report and from screening records from the enrolling audiology clinic.

2. **Age at Diagnosis (months)** was based on the day of the conclusive audiologic assessment that determined individual ear, frequency specific thresholds, and type of hearing loss. This information was generally not obtained at the first audiologic visit and therefore should not be compared directly to the benchmark of three months for beginning audiologic evaluation given by the JCIH.

3. **Age at Fitting of Amplification (months)** was based on the date when the first hearing aids were issued. In many cases, the aids were those purchased or issued, but if loaner instruments were fitted initially, the date of loaner fitting was noted.

4. **Delay of Fitting (months)** was calculated as the difference between age of diagnosis and age of fitting.

5. **Age of Intervention (months)** was based on the first day of actual intervention, determined through a variety of means and converted to age in months. An educational inventory was solicited from interventionists, and these forms recorded beginning intervention dates. These dates were confirmed by parent interview, and in some cases by examination of the Individual Family Service Plan (IFSP). Audiologists in California, by regulation, refer all infants identified with hearing loss to a central office of the California Early Start Program in Sacramento within 48 hours. This office will determine the appropriate local provider of intervention services.

6. **Delay of Intervention (months)** was calculated as the difference between age of diagnosis and age of intervention.

7. **Level of Hearing Loss (dB HL)** was based on hearing threshold information gathered longitudinally from audiology records. For this study, pure tone thresholds for 0.5, 1.0, 2.0, and 4.0 kHz were taken from the most recent or most reliable audiometric report, and the average in the better hearing ear is reported in dB HL. None of the children were noted to have a significant progression in hearing loss during the follow-up period.

**RESULTS**

Of the 63 subjects in the study, 46 were screened during the neonatal period. Of these, 39 failed the screening, and 7 passed. The remaining 17 subjects were not screened. A summary of the group data on age of benchmarks in months can be found on Table 1. A graphic representation of the benchmark ages is shown in the box and whisker plots in Figure 1. Data from infants who passed and failed the newborn hearing screening are plotted separately along with the data from infants not screened. All benchmarks of age at diagnosis, age at fitting of amplification, and age at intervention occur earlier for infants who failed NHS.

Figure 2 displays the data for the computed delays incurred for fitting of amplification and for starting early intervention. These represent the time after diagnosis. During the period of this study, 2002 through 2006, the
median delay between diagnosis of hearing loss and fitting of amplification ranged across screening groups from 1.34 to 1.94 months, and intervention delay ranges from 1.38 to 1.94 months. The negative values for delay of intervention were found when children were enrolled in speech or language intervention before final confirmation of hearing loss. The delays do not change substantially with the screening conditions; rather, hearing aids were fitted and intervention initiated soon after diagnosis if the child had previously failed screening. This delay in fitting of amplification is much less than was noted by Harrison and Roush (1996), who reported a delay of 6–10 months between diagnosis and fitting.

The age at which benchmarks including (1) diagnosis, (2) hearing aid fitting, and (3) intervention, as well as the delay (in months) in hearing aid fitting and intervention, were achieved were compared in infants who were screened and those who were not screened. T-tests were used to determine the effect of screening outcome (pass/fail) on benchmarks, and analyses were performed to compare benchmarks on infants passing NHS with those not screened. Results of t-tests are shown in Table 2.

The children who were screened at birth demonstrated significantly lower ages for diagnosis, hearing aid fitting, and intervention than those who were not screened. However, delays in fitting of amplification and initiation of intervention postdiagnosis are completely independent of screening status in all comparisons. As expected, significant differences were also found in the benchmarks of screened children based on the NHS outcome. Children who failed NHS achieved benchmarks at an earlier age than those who passed. The individual cases of the seven children with hearing loss who passed the neonatal hearing screen will be reviewed in the discussion section.

Finally, the infants with hearing loss who passed NHS achieved benchmarks of age at diagnosis, age at fit, and age at intervention, slightly, but not significantly, earlier than did the infants who were not screened.

Figure 3 plots the age of diagnosis by the age of fitting of amplification for all individuals in the study. The mean data point for each of the subgroups (screened total, screened pass, screened fail, and not screened) is also plotted with filled symbols. The clear advantage of failed screening in terms of early age of diagnosis and fitting is obvious.

The level of hearing loss in the better ear was also compared by screening group, and results are shown on Figure 4 with results of statistical analysis in Table 2. Each of the groups was comprised of children with a wide range of hearing loss. Although the group of screened children who passed appears to demonstrate less hearing loss overall, the differences with other groups were not significant.

Figure 5 displays the relationship between the age at diagnosis and the average hearing loss by screening status. For the infants who failed screening, the age of diagnosis is independent of level of hearing loss. However, for infants not screened and those who passed newborn hearing screening, there is an inverse relationship between the level of hearing loss and the age of diagnosis.

<table>
<thead>
<tr>
<th>Table 1. Median, Mean, and Standard Deviation of Benchmarks in Months Based on Screening Status</th>
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<tr>
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<td></td>
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<tr>
<td>Age at Diagnosis</td>
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<td></td>
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<td>Age of Fitting</td>
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<td>Delay of Fitting</td>
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<td>Age at Intervention</td>
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<td>Delay of Intervention</td>
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**DISCUSSION**

As expected, infants who were screened for hearing loss at birth were diagnosed, fitted with amplification, and entered into early intervention at substantially younger ages than infants who were not screened. The advantage to infants screened for hearing loss over their unscreened counterparts for diagnosis (difference in median age) is 24.8 months, for fitting of amplification is 23.6 months, and for enrollment in intervention is 19.9 months. This significant advantage is seen even when infants who pass the screening are included in the analysis. Also notable is the fact that those infants who are eventually diagnosed with significant hearing loss after passing NHS are found to be delayed in all benchmarks. It is of interest that there is a nonsignificant trend, however, for earlier benchmarks in children who passed NHS than those who were never screened. Certainly with only seven infants in this group, the study may have been underpowered to reach significance. Parents whose infants have passed the screening process in California receive a standard brochure, along with verbal guidance to continue to monitor their child’s
auditory and communication skills and to alert their primary care physicians if any questions arise. It could be argued that families who have been part of the screening process may have been better educated and prepared to watch for hearing loss at a later date and then seek intervention than those who have not been screened. More data are needed to determine if the screening process itself influences outcomes, even when it does not identify hearing loss at birth.

The median age of diagnosis for infants who were screened is 3.03 months, which is in line with the guideline of three months set by JCIH in 2000 and similar to the age of diagnosis given for screened infants in New York (Dalzell et al, 2000). However, if one considers only those infants who failed NHS in this study, the median age is 2.40 months which is within the guideline and similar to the ten weeks reported by Uus and Bamford (2006) in England.

The age of diagnosis for the children who were not screened is 27.83 months. This is later than expected and even later than the age of diagnosis reported by Harrison and Roush (1996) for infants without risk who were diagnosed with mild to moderate hearing loss (22 months). The age of diagnosis for children in this study was, however, similar to that revealed by Mace et al (1991), who found children with hearing loss were identified at 2.1 years.

The fact that the age of diagnosis for unscreened children in this study is as late or later than was reported in the 1990s demonstrates that the advantages in benchmark achievement seen in the screened infants cannot be attributed to a general improvement in awareness regarding the importance of early intervention. Without screening, no improvement in age of identification has been seen since 1991. It is possible that the children born in hospitals that were not screening for hearing loss lived in more rural areas that were later to initiate screening. To investigate this possibility, the socioeconomic status (SES) of the infants in the screened and unscreened groups, as determined by the number of years of the mother’s education, was compared. The mean SES of the unscreened group was
slightly lower (13.64 years) than the screened group (15.51 years), but the difference was not statistically significant ($t = -1.91, P = 0.062, df = 53$). Other between-group factors that might have influenced benchmark achievement, such as degree of hearing loss, have also been ruled out (Table 2).

The JCIH benchmark for fitting of amplification calls for fitting within one month of diagnosis. The range of median “delay of fitting” for this study is between 1.35 and 1.94 months, and screening outcome does not influence the delay. While these results are slightly outside the ideal benchmark, they are encouraging given the reality of delays incurred by families and agencies in obtaining amplification. Our data are in line with those from Uus and Bamford (2006) from Britain who recorded a median fitting delay of five weeks but are better than the 4.5 months reported in New York (Dalzell et al, 2000) or the three to six months reported by Harrison and Roush (1996). It should be noted that agencies in Los Angeles, when possible, fit children with loaner hearing aids while the process of obtaining aids through third party agencies is managed. The date of the first fitting of

Table 2. Results of t-test Comparing Benchmarks across Screening Conditions

<table>
<thead>
<tr>
<th>Screening Status</th>
<th>Diagnosis Age</th>
<th>Fitting Age</th>
<th>Delay of Fitting</th>
<th>Intervention Age</th>
<th>Delay of Intervention</th>
<th>Average Loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>Screened/Not</td>
<td>5.43</td>
<td>5.36</td>
<td>0.37</td>
<td>5.75</td>
<td>1.04</td>
<td>1.039</td>
</tr>
<tr>
<td>P</td>
<td>&lt;0.001</td>
<td>&lt;0.001</td>
<td>0.71</td>
<td>&lt;0.001</td>
<td>0.3041</td>
<td>0.2788</td>
</tr>
<tr>
<td>df</td>
<td>61</td>
<td>61</td>
<td>61</td>
<td>49</td>
<td>49</td>
<td>58</td>
</tr>
<tr>
<td>Pass/Failed</td>
<td>4.78</td>
<td>4.69</td>
<td>0.87</td>
<td>3.12</td>
<td>0.86</td>
<td>0.02</td>
</tr>
<tr>
<td>P</td>
<td>0.001</td>
<td>&lt;0.001</td>
<td>0.38</td>
<td>0.0030</td>
<td>0.394</td>
<td>0.9800</td>
</tr>
<tr>
<td>df</td>
<td>44</td>
<td>44</td>
<td>44</td>
<td>36</td>
<td>36</td>
<td>41</td>
</tr>
<tr>
<td>Passed/Not</td>
<td>0.59</td>
<td>0.49</td>
<td>0.34</td>
<td>1.08</td>
<td>1.31</td>
<td>1.54</td>
</tr>
<tr>
<td>Screened</td>
<td>0.5633</td>
<td>0.6282</td>
<td>0.7333</td>
<td>0.2948</td>
<td>0.2122</td>
<td>0.1388</td>
</tr>
<tr>
<td>df</td>
<td>22</td>
<td>22</td>
<td>22</td>
<td>14</td>
<td>14</td>
<td>20</td>
</tr>
</tbody>
</table>

Note: Comparisons reaching statistical significance are shaded.

Figure 3. Data from individual subjects (open symbols) for age at diagnosis by age at fitting grouped by screening status. Filled symbols with reference lines represent the mean data for the groups.

Figure 4. Box and whisker plot of level of hearing loss in dB by screening status. Group differences were nonsignificant.

Figure 5. Individual subjects’ average hearing level in the better ear is plotted with regression lines by the age of diagnosis for infants who failed screening, passed screening, or who were not screened at birth.
“appropriate” amplification was recorded as the date of amplification fitting regardless of the method by which the aid was obtained.

The median age of intervention for screened infants in this study (10.58 months) is also outside the ideal, six-month benchmark set by JCIIH (2000) and also well above the “age of referral” reported by Dalzell et al. (2000) of five months or Uus and Bamford (2006) of ten weeks. The British study, however, does not make the distinction between referral and actual program implementation. Uus and Bamford mention that the low age of enrollment can be explained by the requirement that infants be referred to early intervention within one day of diagnosis giving the clear impressions that they are reporting referral alone. California regulations also require referral within 48 hours after diagnosis, which is generally accomplished. However, the dates reported in this study are the actual dates on which services were initiated. The age at enrollment was necessarily delayed by the educational process, including the development of the individualized family service or educational plan. The median delay in intervention, following diagnosis, is between 2.46 and 4.12 months, which appears reasonable given the processes that take place.

Influence of Degree of Hearing Loss

The results depicted on Figure 5 demonstrate that, for children who have been identified with hearing loss due to a failed screening, the average level of hearing loss does not influence the age of diagnosis. It should be noted that mild hearing losses were identified by the screening process as well as all other degrees of loss. Children who are either not screened or pass newborn screening are typically identified by the observations of family members and caregivers. Consequently, the negative correlation between level of loss and age of identification, as noted in other studies (Mace et al, 1991; Harrison and Roush 1996), is again seen in these data.

Children with Hearing Loss Who Passed NHS

As mentioned, seven children enrolled in this study had previously passed NHS. The technology used to screen each child is unknown. A summary of those children can be found in Table 3. Of the seven, four have at least one hearing threshold of 30 dB or less in each ear. Three of these four have confirmed or highly suspicious genetic causes for the hearing loss. Subject 28 was confirmed positive for GJB2 (CX-26) mutation, and subjects 133 and 156 are siblings that were not positive for CX-26.

It is certainly possible to pass NHS, performed with either otoacoustic emissions or ABR, if hearing thresholds between 500 and 4000 Hz are below about 35 dB HL. Norton et al (2000) found that all of the current screening techniques, including transient or distortion-product otoacoustic emission or automated ABR, performed very well at identifying hearing loss of 30 dB HL or greater, but performance of all techniques fell off for lesser degrees of hearing loss. The data of Norton et al were based on an ABR screening using a 30 dB nHL stimulus whereas most clinical systems using automated ABR employ a 35 dB nHL click stimulus. It stands to reason that infants with hearing levels better than 35 dB have the possibility of passing the screening regardless of the technology used.

The other interesting question is whether hearing loss is present at birth in all cases of GJB2-related hearing loss. At least two studies have shown that hearing loss symptoms of GJB2 can emerge sometime after the first months of life (Green et al, 2000; Norris et al, 2006). This sample includes only one documented case of GJB2-related hearing loss, and that case demonstrates mild hearing loss. Either the degree of hearing loss or time of onset may explain why this child passed hearing screening.

Three of the children who passed newborn hearing screening, subjects 46, 51, and 130, were later found to demonstrate severe to profound hearing loss in both ears. In two of these cases, infants had a difficult complicated neonatal history including aminoglycocide antibiotics. It must be noted that to pass NHS in California, an infant must pass in both ears. Given the severity of the hearing loss, it is very unlikely that a false negative result occurred in both ears. The most credible explanation for these children’s passing of the NHS is a delay in the onset of the hearing loss.

Outliers: Infants Who Failed NHS and Were Identified Late

In contrast to subjects described above, some of the infants in the study failed NHS and yet were late to be identified. Subject 41 was screened for hearing loss in a hospital that had not yet been certified by the California Children’s Medical Services to participate in the state-wide program. At that time, the hospital did not have procedures in place for informing the parents of the outcome of the screening or for making the appropriate referrals for follow-up testing. Consequently, the parents were not informed of the test outcome, and the child was finally identified by the mother at 19 months of age.

Subject 165 was a premature infant of 30 weeks gestational age who failed an ABR test in the NICU. This child was also identified before the regulations from the state for follow-up testing were in place. The parents took the infant for audiologic evaluations but were told that the baby had passed the audiologic evaluation. This child has a high-frequency sloping hearing loss with low frequencies in the mild to moderate range and high frequencies in the severe range. The current standard for audiologic testing in California calls for frequency specific electrophysiologic testing using tone burst ABR or ASSR. It is recognized that, in the past, many audiologists were using a click response to predict high-
frequency sensitivity. The click stimulus has energy in a broad range of frequencies, and the ABR to a click can be generated by low-frequency regions of the cochlea, especially if those are the areas of greatest sensitivity. In this child, using the once popular but now discredited test strategy of testing with a low-frequency tone burst and a click, ABR testing would have shown hearing to be in the mild range. If audiologic technique is not maximized and large correction factors are used for predicting threshold, the outcome of this test could have been misread by audiologists as normal hearing. Only later, after this child had been enrolled in intensive speech therapy for speech and language delays, did the parents find an audiologic center that used a combination of behavioral measures and frequency-specific electrophysiologic techniques to achieve an accurate diagnosis, but this was when the child was four years and ten months of age!

These cases make it clear that screening alone is not the answer. In the two instances above, the screening was performed and the screening result was appropriate and yet the diagnosis was delayed. The program in California has rigid standards in many areas including standards for inpatient screening and monitored certification of hospitals involved in screening, standards for quality of audiologic care and recommended test protocols, strict standards and monitoring of compliance with follow-up appointments, facilitation of referral to and enrollment in early intervention, and many other high-quality standards for care. The mistakes that led to late diagnosis and intervention are not often seen in the hospitals certified by the Department of Health Services, and indeed, policies have been established to guard against such mistakes.

REFERENCES


Table 3. Characteristics of Children with Hearing Loss Who Passed Newborn Hearing Screening

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<thead>
<tr>
<th>Subject #</th>
<th>Sex</th>
<th>Right Ear</th>
<th>Left Ear</th>
<th>Other factors</th>
<th>DX age</th>
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<tbody>
<tr>
<td>28</td>
<td>M</td>
<td>25</td>
<td>45</td>
<td>CX-26 Positive</td>
<td>21.4</td>
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<tr>
<td>29</td>
<td>M</td>
<td>30</td>
<td>50</td>
<td>20</td>
<td>36.7</td>
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<tr>
<td>46</td>
<td>M</td>
<td>65</td>
<td>75</td>
<td>NICU heart surgery, Ototoxic medication</td>
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<tr>
<td>51</td>
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<td>85</td>
<td>95</td>
<td>Torticolis</td>
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<tr>
<td>130</td>
<td>F</td>
<td>80</td>
<td>95</td>
<td>Premature, Ototoxic meds, Ventilation</td>
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<tr>
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<td>40</td>
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<tr>
<td>156</td>
<td>M</td>
<td>20</td>
<td>30</td>
<td>Not CX-26 or 30</td>
<td>41.1</td>
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