Newborn Hearing Screening: The Great Omission

Albert L. Mehl, MD*‡, and Vickie Thomson, MA§

ABSTRACT. Objective. The advent of technologic improvements in assessing the hearing of newborn infants has made possible the implementation of universal newborn hearing screening. Furthermore, selective screening based on high-risk criteria fails to detect half of all infants with congenital hearing loss. Although universal screening has been recommended by the National Institutes of Health and the Joint Committee on Infant Hearing Screening, data to support this recommendation have been incomplete, and the recommendation has been seen as without solid foundation by many in the pediatrics field. This study was designed to assess the feasibility, accuracy, and cost-effectiveness of a hospital-based hearing screening program for all newborns.

Methodology. Between 1992 and 1996, hospitals in Colorado with 100 or more births per year were targeted to participate in universal hearing screening of newborns. To date, 26 of 52 targeted hospitals, ranging in size from 40 to 3500 births per year, have implemented universal screening. A total of 41 796 infants were screened between 1992 and 1996. Screening was performed using automated auditory brainstem response, otocoustic emission testing, or conventional auditory brainstem response, with follow-up testing performed on those infants who failed initial screening.

Results. Of 41 796 infants screened at birth, 2709 failed initial screening, and of 1296 who have completed reevaluation, 94 have been identified with congenital sensorineural hearing loss (75 bilateral) and an additional 32 identified with conductive hearing loss (14 bilateral). The frequency of bilateral congenital hearing loss requiring amplification therefore is shown to be at least 1 in every 500 newborns. During the study period, an additional 17 children with significant hearing loss not identified until ≥18 months of age were reported voluntarily; all 17 had been born at hospitals not participating in newborn hearing screening.

The false-positive rate for the screening program to date in Colorado is calculated to be 6%, but evolving technology has resulted in improvements to as low as 2%. Positive predictive value of an abnormal screen result is shown to be at least 5%, and as high as 19%, with improving technology. The sensitivity of newborn screening is demonstrated to be at or near 100%. Costs of screening are compared with other screened congenital diseases; although the true cost per child for newborn hearing screening is significantly higher than screening tests performed on blood, the much higher incidence of congenital hearing loss results in a comparable cost per case diagnosed when compared with hypothyroidism or phenylketonuria, for example. The feasibility of early intervention is demonstrated, with amplification by the use of hearing aids being the catalyst for effective treatment.

Finally, the costs of screening and early intervention are compared with the monetary savings in avoiding delayed and therefore intensive therapy and intervention for children not diagnosed at birth. The true cost of screening for one newborn is shown to be between $18 and $33, with an average cost of $25 per infant. The cost per case of congenital hearing loss diagnosed is ~$9600. A model for cost predictions and subsequent intervention savings is presented, and recovery of all screening costs is demonstrated after only 10 years of universal screening in Colorado.

Conclusions. Universal newborn hearing screening is feasible, beneficial, and justified, as indicated by the frequency of the disease, the accuracy of screening tests, the ability to provide early intervention, the improved outcomes attributable to early amplification, and the recovery of all screening costs in the prevention of future intervention costs. Furthermore, the incidence of bilateral congenital hearing loss is alarming, and is, in fact, many times greater than the combined incidence of all newborn screening tests currently performed on blood samples. The demonstrated effectiveness of newborn hearing screening and the availability of early amplification and intervention support the expanding recommendation that every newborn be screened for congenital hearing loss.

Pediatrics 1998;101(1). URL: http://www.pediatrics.org/cgi/content/full/101/1/e4; newborn, congenital, sensorineural, hearing loss, deafness.

ABBREVIATIONS. A-ABR, automated auditory brainstem response; OAE, evoked otoacoustic emission; ABR, auditory brainstem response.

The Committee on Genetics of the American Academy of Pediatrics recently published a comparison of newborn screening programs in the United States and its territories. The list and the accompanying fact sheets about each disease, although comprehensive in addressing screening tests performed on blood, ignore today’s greatest opportunity in newborn screening.

Imagine that there exists a congenital disorder that is detectable at birth with today’s technology, but on average is not diagnosed until age 2½ years, even by experienced physicians; imagine that this same condition can be treated with early intervention to prevent developmental delays and measurable cognitive deficits; imagine that the cost to identify each new case is comparable with the cost of identifying...
Newborn hearing screening has been advocated and performed in the United States since the pioneer work of Marion Downs in 1964. However, the time requirements, variable state of newborn arousal, and subjectiveness of behavioral measurements in the past have prevented practical widespread screening. Parents, although occasionally extremely observant, typically fail to adequately identify hearing impairment in their own children before the first birthday. Even when subsequently proven to be correct, initial parental concerns are frequently discounted by well-
meaning physicians or other health professionals. Currently, the average age of diagnosis for congenital hearing loss is 2 ½ years of age for children not screened at birth, and no trend toward improvement has been observed in this group. Since the inception of the Colorado Newborn Hearing Screening Project, 17 children with hearing loss identified at ≥18 months of age have been reported voluntarily; of these, all were born at hospitals not participating in universal hearing screening.

The incidence of congenital hearing loss is shown from the Colorado experience to be alarmingly high, with bilateral hearing loss present in at least 1 of every 500 newborns. For comparison, the incidence of commonly screened newborn disorders would be galactosemia, 2 per 100 000 births; phenylketonuria, 10 per 100 000 births; hypothyroidism, 25 per 100 000 births; and bilateral sensorineural hearing loss, 200 per 100 000 births (Table 1).

Before the availability of automated techniques for screening, the use of traditional ABR was performed selectively in some hospitals, with a high-risk registry used as a tool for selecting those infants who would be tested. Although the implementation of high-risk guidelines has been extremely variable in the past, a thorough screening for high-risk categories would include assessment of each of the following areas: asphyxia, meningitis, congenital or perinatal infections, anatomic defects or stigmata, hyperbilirubinemia, family history of hearing loss, low birth weight, ototoxic medications, and neonatal illnesses requiring mechanical ventilation.1 When Colorado infants with proven congenital hearing loss were reviewed retrospectively for any of these high-risk criteria, 63 of 126 affected newborns, or 50%, were shown to have no risk factors, and therefore would have never received newborn hearing screening in the traditional model of screening only at-risk infants.

In place of standard ABR testing, two newer techniques for universal neonatal screening are currently available, and both methods were used in the various Colorado hospitals studied. A-ABR allows for computerized interpretation and pass–fail reporting. This computerization makes it possible to perform screening with personnel of variable backgrounds and training. In Colorado, ~80% of all newborn screening is, in fact, performed by volunteers, technicians, or nurses, rather than by audiologists. A-ABR is also the preferred test in a setting of early hospital discharge, because neither middle ear fluid nor ear canal debris, often still present in the first 12 to 24 hours of life, will adversely affect the result. A-ABR requires infants to be asleep or in a quiet state at the time of testing.

OAE is also considered an appropriate screening tool. Similarly performed on a sleeping or quiet newborn, the test is quick in producing a result and involves less cost for disposable items. Interpretation by an audiologist is required, however, making this technique less appealing for smaller hospitals. The test is not accurate when the middle ear has not yet been adequately aerated after birth or when debris remains in the ear canal.

### TABLE 1. Summary of Selected Colorado Newborn Screening Programs: A Comparison of Existing Screening Programs and Proposed Universal Infant Hearing Screening

<table>
<thead>
<tr>
<th></th>
<th>Congenital Hearing Loss</th>
<th>Hypothyroidism</th>
<th>Phenylketonuria</th>
<th>Cystic Fibrosis</th>
<th>Hemoglobinopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frequency per 100 000 births</td>
<td>260 (200 Bilateral)</td>
<td>25</td>
<td>7</td>
<td>50</td>
<td>13</td>
</tr>
<tr>
<td>Yearly Colorado births</td>
<td>54 000</td>
<td>54 000</td>
<td>54 000</td>
<td>54 000</td>
<td>54 000</td>
</tr>
<tr>
<td>Number positive, first screen</td>
<td>3500 (1620 Expected)</td>
<td>600</td>
<td>5</td>
<td>600</td>
<td>600</td>
</tr>
<tr>
<td>Number of children diagnosed</td>
<td>140 (110 Bilateral)</td>
<td>15</td>
<td>4</td>
<td>27</td>
<td>7</td>
</tr>
<tr>
<td>Positive predictive value</td>
<td>5% (19% Expected)</td>
<td>3%</td>
<td>80%</td>
<td>4%</td>
<td>1%</td>
</tr>
<tr>
<td>Average age of diagnosis if unscreened</td>
<td>30 months</td>
<td>3–12 Months</td>
<td>3–12 Months</td>
<td>42 Months</td>
<td>3–36 Months</td>
</tr>
<tr>
<td>Cost of initial screen per child</td>
<td>$25</td>
<td>$3</td>
<td>$3</td>
<td>$3</td>
<td>$3</td>
</tr>
<tr>
<td>Screening cost per confirmed diagnosis</td>
<td>$9600 ($12 300 Bilateral)</td>
<td>$10 800</td>
<td>$40 500</td>
<td>$6000</td>
<td>$23 100</td>
</tr>
<tr>
<td>Effectiveness of treatment</td>
<td>2+</td>
<td>3+</td>
<td>3+</td>
<td>1+</td>
<td>2+</td>
</tr>
<tr>
<td>Clinical result of delayed diagnosis</td>
<td>Language delay, academic delay, psychosocial difficulties, cognitive delays</td>
<td>Lethargy, confusion, poor memory, myxedema, coma, cretinism</td>
<td>Seizures, tremors, severe irreversible mental retardation</td>
<td>Malnutrition</td>
<td>Bacterial sepsis, anemia, sickling crisis</td>
</tr>
</tbody>
</table>

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In addition, the two tests have been combined in some centers, effectively creating a two-stage screening process while infants are still in the hospital. To achieve effective universal screening, initial screening tests are best performed before newborn discharge from the hospital; screening tests performed after hospital discharge can be influenced negatively by the failure of families to return for retesting and by the failure of newborns to be in a quiet state at their appointed time for rescreening.

Newborns who fail initial screening must return for adequate follow-up testing, and appropriate systems need to be in place to ensure timely recall. Although a number of Colorado newborns who failed initial screening have follow-up evaluation in progress, there is clearly an opportunity for improvement in ensuring retesting of all newborns who fail the initial screening. Initial follow-up testing typically includes a rescreening by one or more of the techniques described above. Infants who fail this second screening require a comprehensive audiology evaluation.

Of 41,796 newborns screened to date in Colorado, no evidence of even a single false-negative test result has been discovered; the sensitivity of newborn screening is therefore at or near 100%. Since the inception of newborn hearing screening in Colorado, the cumulative false-positive rate is ~6%. With improving technology, a trend toward greater specificity is noted; specifically, the second generation of A-ABR equipment currently used widely in Colorado (Algo 2, Natus Medical Inc) has allowed for a false-positive rate as low as 2%.

Even this lower rate of 2%, when compared with other commonly screened newborn disorders, may appear to be higher than desirable. It is important, however, to interpret this rate carefully in the context of a disease that is many times more common than other conditions screened. A more meaningful number, therefore, is the positive predictive value of the test, namely the number of true positive results divided by the total number of positive screening tests. The positive predictive value for all newborn hearing screening to date in Colorado is \( \approx 5\% \); calculated for second-generation A-ABR equipment only, the positive predictive value is now as high as 19%. This compares favorably in Colorado with the positive predictive values for hemoglobinopathy screening (1%), cystic fibrosis screening (4%), and hypothyroidism screening (3%). In fact, the positive predictive value for newborn hearing screening is exceeded in Colorado only by screening for phenylketonuria (80%) (Table 1).

Unilateral congenital hearing loss is, admittedly, a relatively minor disability in language and cognitive development. It is, nevertheless, an educational disability that is managed easily, but only after adequate and, preferably, early diagnosis.

Bilateral congenital hearing loss, however, is of greater significance. On average, language delay at the time of diagnosis ranges from 12 months to \( \approx 2 \) years. Treatment after diagnosis, typically after age 2, requires intensive therapy for both articulation and expressive–receptive language development. Even years of intensive intervention will fall short of achieving results comparable with language development in children with normal hearing.

With neonatal screening and early diagnostic confirmation, amplification and intervention can be initiated within the first 3 months of life. Because only 10% of affected newborns have profound hearing loss (7 of 75 newborns with bilateral sensorineural hearing loss identified to date in Colorado), major improvements in outcome can be obtained with amplification, careful audiology follow-up, and appropriate habilitation. With early amplification and appropriate therapy, children who are hearing-impaired are capable of achieving normal speech–language developmental milestones. Yoshinaga-Itano and coworkers have shown that a cohort of affected newborns identified through newborn screening achieved near-normal language development, significantly exceeding the language development of comparable children not identified until after 6 months of age.\(^5\)

Early consistent amplification is the catalyst for effective intervention for newborns with sensorineural hearing loss as well as for infants with conductive hearing loss awaiting later surgical intervention. Even though the 10% of hearing-impaired newborns with profound hearing loss receive limited benefit from traditional amplification, newer technologies may be introduced to facilitate communication development and to offer families a range of options for their child’s education. In addition, surgical innovations such as cochlear implants are increasingly available. In the case of profound hearing loss, use of sign language may be initiated in early infancy to facilitate communication.

For all infants affected, management by a certified audiologist is necessary, in conjunction with a primary care physician invested in coordinating adequate follow-up. Evaluation by a language specialist is essential, although some amplified infants require only limited formal therapy. Individual detection of newborns affected, combined with family history and additional medical evaluation when indicated, also allows for genetic counseling and recurrence risk estimation when appropriate.

It is important to note that a normal screening test does not preclude the subsequent diagnosis of an acquired sensorineural hearing loss (secondary to meningitis, for example), nor does it preclude a progressive hearing loss with onset later in childhood. Furthermore, the possibility of subsequently acquired conductive hearing loss (from chronic middle ear effusion, for example) will, of course, require the clinician to continue a vigilant approach to assessment of hearing acuity and language development, even when newborn screening has been documented as normal.

Screening for congenital hearing loss is clearly a significant undertaking. Because this screening does not involve a blood test, the costs are independent of structures already in place to support universal blood screening tests. The true cost for each infant screened is estimated to be $25 per infant, including labor costs, disposable supplies, and amortized cap-
diagnosis of bilateral sensorineural hearing loss, with zero educational savings for treatment of unilateral hearing loss. (Costs and system, for the first year and each subsequent year; finally, assuming all educational savings apply only to prevention of delayed birth would allow for a one-level shift to less-intensive educational setting for only half of each cohort as they enter the educational ($2300 per year excess costs), and 34% consultative/itinerant programs ($700 per year excess costs), and assuming that identification at exceeding standard education by $25 000 per year), 23% self-contained classroom ($8300 per year excess costs), 28% resource programs of early intervention preschool home program ($2600 per year), followed by 1 year of specialized preschool ($6200 per year), avoidable prevention of delayed diagnosis of bilateral sensorineural hearing loss; zero therapy savings estimated if unilateral.  

response with sedation.

all newborns failing initial screening.

screening costs.

Notes: Calculations based on the following assumptions:

- All cost figures reflect estimated true costs, not charges.
- 54 000 births per year, every year.
- Frequency of congenital bilateral hearing loss 2 per 1000 newborns
- Cost per initial screening, $25 per infant; cost of follow-up program and coordinator, estimated at $100 000 per year included in initial screening costs.
- Cost of confirmatory evaluation, $125 per infant who fails initial screening, unilateral or bilateral; costs based on a refer rate of 3.0% of all newborns failing initial screening.
- Cost of intervention associated with amplification $1200 per year for 2 years in follow-up costs. The cost of the amplification device, required both for infants with early diagnosis and with late diagnosis, has been excluded from calculations.
- Age of diagnosis if not screened at birth assumed to be 30 months.
- Cost of confirmatory late diagnosis estimated to be $400 per affected child, for behavioral testing and brainstem auditory evoked response with sedation.
- Cost of therapy savings estimated at $40 true cost per session, 3 days per week, 50 weeks per year, for 3 years, for each child with prevention of delayed diagnosis of bilateral sensorineural hearing loss; zero therapy savings estimated if unilateral.
- Cost of preschool educational savings estimated as follows: assuming that half of children with delayed diagnosis would require 1 year of early intervention preschool home program ($2600 per year), followed by 1 year of specialized preschool ($6200 per year), avoidable if diagnosed at birth.
- Cost of school-age educational savings estimated as follows: Assuming delayed diagnosis results in 13% residential placement (cost exceeding standard education by $25 000 per year), 23% self-contained classroom ($8300 per year excess costs), 28% resource programs ($2300 per year excess costs), and 34% consultative/itinerant programs ($700 per year excess costs), and assuming that identification at birth would allow for a one-level shift to less-intensive educational setting for only half of each cohort as they enter the educational system, for the first year and each subsequent year; finally, assuming all educational savings apply only to prevention of delayed diagnosis of bilateral sensorineural hearing loss, with zero educational savings for treatment of unilateral hearing loss. (Costs and percentages: Colorado Department of Education, 1993 data.)
- Model assumes that infants identified with congenital sensorineural hearing loss do not move out of Colorado during the 10-year period.
- Negative net costs shown in parentheses are the equivalent of net savings.

The cost of screening for congenital sensorineural hearing loss, however, must again be interpreted in the context of a disorder that is by no means rare. In Colorado, the screening costs required to identify correctly one new case of congenital hearing loss are calculated to be $9600. Even though the cost per test is much less for each blood test performed, the newborn genetic screen diseases are much less common. The cost per case diagnosed, therefore, is similar: $10 000 per case for hypothyroidism, $23 000 per case for hemoglobinopathy, and $40 000 per case for phenylketonuria (Table 1). Finally, the labor costs of using the traditional high-risk registry followed by selective screening to identify children with hearing loss can be expected to approach the labor costs of universal screening. However, a thorough application of this traditional high-risk method would fail to detect 50% of newborns affected.

In 1997, the state of Colorado has legislated a requirement for more complete penetration of universal newborn hearing screening among the state’s hospitals; the total estimated cost to screen every one of Colorado’s 54 000 births each year, followed by confirmatory testing for those who failed the initial screen, would be $1 650 000. Follow-up associated with amplification of infants affected would require another $260 000 per year on average. For the first 2 ½ years, no savings would be expected from a universal screening program, because unscreened hear-

<table>
<thead>
<tr>
<th>Year</th>
<th>Initial Screen Costs</th>
<th>Confirmatory Evaluation Costs</th>
<th>Cost of Intervention</th>
<th>Cumulative Cost</th>
<th>Cumulative Eval and Therapy Savings</th>
<th>Cumulative Education Savings</th>
<th>Net Cost or (Savings)</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>1.45</td>
<td>0.2</td>
<td>0.07</td>
<td>1.72</td>
<td>0</td>
<td>0</td>
<td>1.72</td>
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<tr>
<td>2</td>
<td>1.45</td>
<td>0.2</td>
<td>0.20</td>
<td>3.56</td>
<td>0</td>
<td>0</td>
<td>3.56</td>
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<td>3</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>5.47</td>
<td>0.10</td>
<td>0</td>
<td>5.37</td>
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<td>4</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>7.38</td>
<td>0.79</td>
<td>0.14</td>
<td>6.46</td>
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<td>5</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>9.29</td>
<td>2.13</td>
<td>0.61</td>
<td>8.65</td>
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<tr>
<td>6</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>11.2</td>
<td>4.04</td>
<td>1.57</td>
<td>5.84</td>
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<td>7</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>13.1</td>
<td>6.03</td>
<td>2.25</td>
<td>4.82</td>
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<td>8</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>15.0</td>
<td>8.01</td>
<td>3.41</td>
<td>5.58</td>
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<td>9</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>16.9</td>
<td>10.0</td>
<td>4.81</td>
<td>2.09</td>
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<td>10</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>18.8</td>
<td>12.0</td>
<td>6.43</td>
<td>0.37</td>
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<tr>
<td>11</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>20.7</td>
<td>14.0</td>
<td>8.27</td>
<td>(1.57)</td>
</tr>
<tr>
<td>12</td>
<td>1.45</td>
<td>0.2</td>
<td>0.26</td>
<td>22.6</td>
<td>16.0</td>
<td>10.4</td>
<td>(3.80)</td>
</tr>
</tbody>
</table>
ing-impaired children are diagnosed, on average, some 30 months later. Nevertheless, beginning in the 3rd year, true savings can be calculated in the avoidable costs of later evaluation and intensive speech–language intervention. Without screening, such intervention is required in hope of achieving an imperfect recovery of language development and cognitive losses. Furthermore, additional expenses accrue for children not diagnosed at birth who go on to require some form of special intervention once they reach school age, ranging from itinerant and resource programs, to self-contained classrooms, to residential placement. If only half of hearing-impaired children realized some ultimate savings in school-based costs because of newborn screening and early amplification, a universal screening program in Colorado could recover all screening costs after only 10 years through subsequent savings in avoided intervention (Table 2). Recovery of all initial costs (and subsequent cost savings) is independent of improved developmental outcomes, a worthy goal in and of itself.

Identification of congenital hearing loss is well recognized as an opportunity for effective screening and early treatment. National Institutes of Health has recommended that newborn hearing screening be implemented universally. The Joint Committee on Infant Hearing, representing the American Academy of Pediatrics, the American Academy of Otolaryngology, the American Academy of Audiology, and the American Speech–Language–Hearing Association, has similarly recommended hearing screening for all newborns.

The pediatrics community as a whole, however, is only beginning to realize the potential of new technology in diagnosing this disorder accurately in a newborn baby. Congenital hearing loss is far too common, and the developmental disabilities far too devastating and expensive to allow the initial costs to be a barrier to pursuing this exciting initiative. The Newborn Hearing Screening Project of the State of Colorado encourages all neonatologists, pediatricians, and family practitioners to include newborn hearing screening as a necessary and effective test for every newborn.

REFERENCES
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