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ABSTRACT. Objective. Although previous studies have documented the feasibility and benefits of universal newborn hearing screening in selected hospitals, none have reviewed the effectiveness of regionally mandated participation of large numbers of hospitals with variable levels of motivation to succeed. The purpose of this study was to measure hospital participation and overall screening success in a statewide program for universal newborn hearing screening and to track improvements in program establishment and outpatient follow-up over time.

Methods. Four Colorado hospitals began voluntarily performing hearing screening before hospital discharge on all newborns in 1992. By 1996, 26 Colorado hospitals were participating in universal newborn hearing screening. The publication of screening results from these early years served as a catalyst for legislation requiring increased hospital participation in establishing universal screening programs. Data systems were subsequently developed to improve statistical tracking and follow-up. Eight years' worth of cumulative study data as well as the results from calendar year 1999 (the year of greatest hospital participation) were reviewed for collective measures of successful screening and follow-up. Three hospitals did not initiate newborn hearing screening programs until after the study period ended in 1999. Of the 57 hospitals that were screening newborns in 1999, the chosen method of screening at 52 hospitals was automated auditory brainstem response testing; 3 hospitals used otocoustic emission testing, and the remaining 2 hospitals used 2-stage screening. Hearing loss was defined as a threshold of 35 decibels or greater in 1 or both ears at the time of confirmatory testing.

Results. During the full 8-year study period, 1992 to 1999, 148,240 newborns were screened. A total of 291 infants who were born during the study period received a diagnosis of congenital hearing loss. In this cohort of 291 children, the cumulative frequency of bilateral hearing loss was 71% (range: 48%–94% by calendar year), the frequency of sensorineural hearing loss was 82% (range: 67%–88%), and the frequency of 1 or more risk factors was 47% (range: 37%–61%). During calendar year 1999, a total of 63,590 births were recorded at 60 birthing hospitals in Colorado. The families of 263 (0.4%) of these newborns refused newborn hearing screening. Of the remaining 63,327 newborns, 87% (55,324 infants) were screened for hearing acuity before hospital discharge, a far greater percentage than the 23% of all newborns screened during the first 5 years of voluntary hospital participation, and approaching the American Academy of Pediatrics's recommendation of 95% of newborns completing hospital-based testing in a successful screening program. As a result of this statewide hearing screening program, congenital hearing loss was diagnosed in 86 Colorado newborns during 1999, representing an occurrence rate of approximately 1 affected child in every 650 newborns. In this group of 86 infants, 59 had bilateral sensorineural hearing loss, 17 had unilateral sensorineural hearing loss, 4 had bilateral conductive hearing loss, and 6 had unilateral conductive hearing loss. Mild hearing loss was present in 6 infants, moderate hearing loss was present in 42 infants, severe hearing loss was present in 33 infants, and profound hearing loss was present in the remaining 5 infants. Only 32 of the 86 affected newborns in 1999 had 1 or more risk factors for hearing loss subsequently identified. After failing an initial hospital-based screening at 1 of the 57 participating hospitals in 1999, 2.3% of infants screened (1283 newborns) were referred for follow-up testing, easily exceeding the standard of <4% recommended by the American Academy of Pediatrics. Similarly, the false-positive rate of 2.2% during 1999 exceeded the recommended standard of <3%. Of the infants who failed their initial screening, 76% (978 infants) had documented follow-up testing to confirm or exclude congenital hearing loss, a percentage significantly improved from a follow-up rate of 48% during the first 5 years of screening, although not yet achieving the standard of 95% recommended by the American Academy of Pediatrics. Nine participating hospitals, however, were able to document appropriate follow-up for 95% or more of the infants who failed their initial screening tests. The median age of diagnosis of congenital hearing loss during 1999 was 2.1 months; 71% of affected infants were identified by 3 months of age (the recommended standard for age of diagnosis), and 92% of affected newborns were identified by 5 months of age. Measures of screening success were compared for large, mid-sized, and small hospitals. Increasing hospital size, as measured by the number of births per year, was associated with an increasing percentage of newborns who were successfully screened. It was notable that smaller hospital size was associated with increased referral rates for follow-up testing, whereas larger hospital size was associated with the highest recapture rate for follow-up testing.

Conclusions. Universal screening for congenital hearing loss is demonstrated to be feasible in a large regional effort of legislatively mandated participation. The success of such an endeavor is dependent on educational efforts for community professionals, commitment on the part of hospital administrators and staff, and effective follow-up services. The program has demonstrated increasing screening success over time, as well as increasing awareness and education for professionals regarding follow-up services.
Congenital hearing loss has been recognized for decades as a serious disability for affected children, with a delay in diagnosis of 2 years or more being the rule rather than the exception. In 1993, the National Institutes of Health recommended that every newborn infant have a hearing test performed in the first few months of life. This recommendation was soon followed by a similar guideline prepared by the Joint Committee on Infant Hearing (representing the American Academy of Pediatrics, the American Academy of Audiology, the American Academy of Otolaryngology, the American Speech-Language-Hearing Association, and directors of state speech and hearing programs), concurring that hearing screening should be performed on every newborn. Many physicians, however, received these guidelines with skepticism. The effort was seen as perhaps overzealous, and the feasibility of mass screening programs was questioned. Furthermore, the efficacy of early intervention was largely unproved, initial costs were substantial if not staggering, and the potential harm of false-positive screening results suggested caution.

Nevertheless, evidence in support of this aggressive universal screening recommendation accumulated, as increasing numbers of hospitals implemented newborn hearing screening programs. In Colorado, results from voluntary participation at 26 hospitals revealed the frequency of newborn hearing loss to be alarmingly high, affecting as many as 1 in every 500 newborns. Furthermore, the earlier Colorado data revealed that, when reviewed retrospectively, at most half of all newborns with congenital hearing loss would have been detected using older high-risk protocols. In addition, the initial costs were shown to be completely recoverable in conservative cost savings models, independent of improved outcome.

With early detection and treatment of an increasing number of children with congenital hearing loss in Colorado, comparative developmental outcomes could be more critically assessed. In research that has subsequently been confirmed by other investigators, Yoshinaga-Itano et al demonstrated the significantly improved outcomes for children who have congenital hearing loss and received early intervention when compared with a cohort of similar children who did not receive the benefit of early screening and detection. Similarly, independent of specific screening protocols and measures of screening follow-up success, affected infants who were born in a hospital with an established screening program had significantly improved outcomes when compared with those who were born in hospitals that did not screen.

More important, the critical window of intervention was shown to be much earlier than previously suspected, with delays in diagnosis of only 6 to 12 months associated with significant and ongoing delays in language development. The disability caused by auditory sensory deprivation during critical brain development, previously understood in animal studies of the auditory system, was now recognized as being more clearly associated with clinical developmental delays in humans.

Finally, false-positive rates have shown gradual but significant improvement, as advances in technology and training methodology evolve. Concurrently, protocols for rescreening before hospital discharge or 2-stage screening using different techniques have improved false-positive rates of 6% to 10% (or greater) to levels as low as 1% to 2%,. In addition, investigators have measured the potential parental distress that results from a false-positive hearing screening test, with reassuring results.

Successful universal newborn hearing screening is now a reality at many motivated hospitals across the United States and throughout the world. Several multiple-hospital systems have published impressive results. Nevertheless, if universal screening is justifiable, then it must also be proved effective and feasible across geographically diverse hospital systems, with variable fiscal structures, reimbursement strategies, staffing philosophies, equipment availability, and motivation to succeed.

Despite Colorado’s initial screening success, the demonstrated program costs and questioned justifications for universal screening resulted in a plateau of hospital participation during 1996. In response to new, increasingly defensible screening recommendations, the Colorado legislature passed in 1997 House Bill 97-1095, promoting statewide expansion of hospital-based newborn hearing screening. Although the legislation has subsequently served as a model for other state legislative efforts, it was clearly an eclectic mixture of medical intervention philosophies and social engineering. With legislative hesitation concerning this (or any) “unfunded mandate,” hospital association concerns about funding and reimbursement structures and physician anxiety about legislating the practice of medicine, the resulting law became an uncomfortable but workable compromise. The resulting “soft mandate” required that voluntary hospital participation be measured against a deadline; if aggregate hospital screening were to fall short of 85% of all Colorado newborns after 2 subsequent years of hospital enrollment in hearing screening participation (July 1999), then the Colorado Board of Health would be directed to promulgate rules requiring universal screening at every birthing hospital. The legislation failed to address reimbursement specifically, and no new funding was provided for tracking mechanisms through the Colorado Department of Public Health and Environment.

This article reports the results of a statewide screening program, from earlier years of voluntary hospital enrollment through later years of legislative
directives. The study represents the first publication of broad population-based hospital screening efforts and measures the relative success of legislation in achieving universal newborn hearing screening.

METHODS

The intent of this study was to measure hospital participation and overall screening success, with a comparison of screening penetration before and after legislative intervention. The period of study included calendar years 1992 through 1999. (Results were tabulated after June 2000, allowing at least 6 months for documentation of follow-up for infants who were born late in calendar year 1999.) Beginning in 1992, 4 Colorado hospitals voluntarily began universal newborn hearing screening pilot programs. Over subsequent years, additional hospitals were encouraged and assisted in their fledging screening efforts by the Colorado Department of Public Health and Environment. Hospital audiologists or their designees were enlisted to provide tracking and statistical reporting to this state entity, and monthly summaries of births and screening success were collected from every birthing hospital. Initial paper reporting mechanisms were gradually expanded and computerized, leading eventually in 1999 to data inclusion within the existing structure of the Colorado electronic birth certificate, currently used by most Colorado hospitals.

All birthing hospitals were included as potential screening locations, and participation by hospital was monitored. Parents at participating hospitals were informed of the availability of newborn hearing screening before hospital discharge, and parental consent for testing was obtained. Educational sessions were provided to train physicians, audiologists, hospital staff, and related personnel. Established by legislation in 1997, the Colorado Infant Hearing Advisory Committee, composed of physicians, audiologists, early interventionists, educators, public health professionals, parent and family advocates, and members of the deaf community, created and published guidelines for effective screening, diagnosis, and intervention.

Congenital hearing loss was defined as hearing thresholds of 35 dB or greater in 1 or both ears, as measured by diagnostic brainstem auditory evoked response testing. Confirmed hearing loss reports were collected from audiologists throughout the state, and the assistance of the “CO-Hear” state audiology consulting network was enlisted to ensure continuing follow-up and reporting.

RESULTS

Hospital participation increased from 4 hospitals in 1992 to a plateau of 27 birthing hospitals in 1995 and 1996. After legislation was passed in 1997, hospital participation rapidly increased to a peak of 57 of 60 birthing hospitals at the end of 1999. Of the remaining 3 hospitals, 2 hospitals (38 births and 437 births, respectively, in 1999) initiated universal newborn hearing screening during calendar year 2000, and the final remaining facility (1146 births in calendar year 1999) began screening all newborns early in 2001 (Fig 1).

Hospital sizes ranged from 2 births per year to 5199 births per year during calendar year 1999. Of 57 participating hospitals in 1999, 52 used automated auditory brainstem response (AABR) technology as their equipment of choice, 3 hospitals used otoacoustic emission (OAE) testing for hospital-based screening, and 2 hospitals elected to perform 2-stage screening, using OAE initially and AABR rescreening before hospital discharge for those infants who did not pass OAE screening.

During the study period, 148 240 newborns were screened and 291 infants were identified as having congenital hearing loss (Fig 2). During 1999, the last and most complete year of screening, there were 63 590 births; as a result of newborn hearing screening, 86 infants subsequently received a diagnosis of confirmed congenital hearing loss, resulting in a prevalence of 135 affected children per 100 000 live births. Excluding the group of 8266 newborns who were not screened before hospital discharge (and therefore presumed to be at risk for later diagnosis of congenital hearing loss), the frequency of congenital hearing loss is calculated to be 155 per 100 000 live births, or approximately 1 in every 650 newborns.

During 1999, specific refusal by parents to participate in newborn hearing screening was documented for 263 newborns (0.4% of those families who were approached). Of the remaining 63 327 infants born at all 60 birthing hospitals, 55 324 (87%) were successfully screened before hospital discharge (Fig 3). For comparison, only 19% of Colorado newborns were screened during the years of voluntary hospital participation, 1992 through 1996.4

Of all infants screened during 1999, 1283 newborns (2.3%) did not pass initial hospital-based screening and were referred for follow-up testing. When compared by method of testing, the combined referral rate for the 52 hospitals that used AABR screening was 1.5% (49 325 screened, 729 referred); the combined referral rate for the 3 hospitals that used OAE screening was 11.0% (1957 screened, 216 referred), and the combined rate for the 2 hospitals that used 2-stage screening was 8.4% (4042 screened, 338 referred). Of the 1283 infants who were referred for follow-up testing, 978 (76%) had documentation of return for outpatient follow-up. For comparison, only 48% of newborns who failed initial screening...
during calendar years 1992 to 1996 had documentation of follow-up.\textsuperscript{4}

Of the group of infants who returned after an abnormal screening test in 1999, 86 infants (9%) had congenital hearing loss subsequently confirmed. In this cohort of 86 deaf or hard-of-hearing newborns, 59 had bilateral sensorineural hearing loss, 17 had unilateral sensorineural hearing loss, 4 had bilateral conductive hearing loss, and 6 had unilateral conductive hearing loss. Mild hearing loss (35–40 dB threshold) was present in 6 of the 86 affected infants; 42 had moderate hearing loss (41–70 dB), 33 had severe hearing loss (71–90 dB), and 5 had profound hearing loss (91 dB or greater). The median age of diagnosis for affected infants who were born in 1999 was 2.1 months. Of the 86 affected children, 29 had confirmatory diagnosis by 1 month of age, an additional 22 by 2 months of age, 10 more by 3 months of age, and an additional 9 by 4 months of age; 9 of the infants had confirmatory diagnosis during the fifth month of life, and the remaining 7 had a diagnosis at 6 months of age or later.

For each calendar year, the percentage of newborns who had a hearing impairment and were identified as having high-risk factors, as defined by the Joint Committee on Infant Hearing,\textsuperscript{18} ranged from 37% to 61%, with a cumulative percentage of 47% (138 of 291 newborns). High-risk factors for congenital hearing loss were identified in only 32 of the 86 affected infants (37%) who were born during 1999.

The majority of affected newborns had bilateral hearing loss; by calendar year, the percentage of newborns who were hard-of-hearing with bilateral hearing loss ranged from 48% to 94%, with a cumulative fraction of 71% (206 of 291 infants). Similarly, the majority of infants who had a hearing impairment (82% cumulatively) had sensorineural hearing loss (239 of 291 newborns), ranging from 67% to 88% by calendar year.

Statistical results for 46 hospitals that were performing hearing screening throughout calendar year 1999 were analyzed further. These hospitals were divided into groups by birthing volume during 1999, with 4 resulting categories: 1 to 400 births (16 hospitals), 401 to 1000 births (8 hospitals), 1001 to 2000 births (11 hospitals) and 2001 to 6000 births (11 hospitals). When compared among these hospital groups, the percentage of newborns who were successfully screened before hospital discharge showed an increasing trend commensurate with increasing hospital size; only 60% of newborns from the smallest hospitals were screened successfully, whereas 92% of infants who were born at the largest hospitals were screened (Fig 4).

For the 46 hospitals that participated throughout calendar year 1999, referral rates for outpatient retesting after failed initial screening were highest for small hospitals, averaging 5.8% of all infants screened. Referral rates were lowest (1.3%) for hospitals with 401 to 1000 births per year, whereas hospitals with >1000 births per year had referral rates of 2.2% (Fig 5). The largest hospitals had the highest rate of successful recapture of infants for rescreening after a failed initial screening; in this hospital grouping, 81% of newborns who did not pass the initial screening had documented return. All groupings of
hospitals that had <2000 births per year had similar documented return rates of 66% to 68% (Fig 6).

DISCUSSION

Screening every newborn for congenital hearing loss is an undertaking of no small measure. Nevertheless, these most recent Colorado data again illustrate the alarming frequency of the condition and the resulting importance of population screening. The recruitment of diverse hospitals of varying size to participate in a statewide effort is demonstrated to be feasible; all 60 birthing hospitals had implemented universal newborn hearing screening programs as of 2001. As evidence mounts to support earlier recommendations for universal hearing screening, hospitals and hospital systems have increasingly begun to question not why to screen all newborns but how to screen all newborns.

In this study, congenital hearing loss is confirmed to be not at all rare, affecting approximately 1 in
every 650 newborns, a frequency far greater than the combined frequencies of all of the metabolic conditions currently recommended for newborn screening. In addition, the study once again demonstrates the futility of using a high-risk registry approach for diagnosing congenital hearing loss. Cumulatively during the 8 years of study, only 47% of infants with confirmed hearing loss had evidence of 1 or more high-risk factors.

These 1999 Colorado statistics show the state to be approaching the recommended screening program standards published by the American Academy of Pediatrics Task Force on Newborn Hearing.19 With a goal of screening all newborns, the task force considered 95% penetration as an indicator of effective screening. In Colorado, of 46 hospitals screening throughout 1999, 23 hospitals had screening rates of 95% or greater and an additional 6 hospitals had screening rates between 90% and 95%. Including 11 hospitals that began screening during calendar year 1999 and 3 hospitals that had not yet initiated screening, the state nevertheless managed to screen 87% of all children who were born during 1999. Increasing hospital size was associated with higher rates of screening penetration, likely representing an economy of scale in the development of programs designed to screen large numbers of infants each day.

The task force recommended that the referral rate for follow-up of abnormal screening results be no greater than 4%. In Colorado during 1999, including all 57 screening hospitals, the cumulative referral rate was 2.3%. Twenty-three hospitals recorded referral rates of <2%, and of these 23, 14 hospitals managed to achieve referral rates of <1%. Referral rates for rescreening were highest among the smallest hospitals; this may be an indication that repetition and familiarity with screening techniques could result in lower referral rates. The combined referral rate for the 3 hospitals that use OAE alone was 11%, similar to rates reported in earlier literature but far above the recommended threshold of 4%. Two-stage screening should have the potential of improving this rate significantly, although the 2 hospitals that use this protocol had mixed success: 1 had a referral rate after 2-stage screening of 0.7%, and the other had a rate of 12.3%.

The task force recommended that, after eliminating from consideration the children with subsequently confirmed hearing loss, the false-positive rate for newborn hearing screening be no greater than 3%. During 1999 in Colorado, 1197 of 55,324 infants screened had a false-positive screening result, for a false-positive rate of 2.2%. This rate compares very favorably to previously published rates of 5% to 10% or more.

In addition to improving technology and experience-based training protocols implemented during the past decade, Colorado’s success in the area of false-positive rates is clearly the result of a decision to emphasize screening with AABR rather than OAE; although OAE may offer other advantages, such as limiting the cost of disposable supplies, a higher reported false-positive rate has led to the recommendation of 2-staged screening before hospital discharge.2,11

Although the AABR and OAE technologies both are accepted as reliable measures for newborn hearing screening, no conclusion can be drawn from this study about the possibility of false-negative testing. However, one category of “false negative” testing should be specifically noted. Auditory neuropathy is a rare but significant disorder whereby the cochlear and external hair cells are intact but the “retro-cochlear” central auditory mechanism fails to receive and/or process auditory impulses adequately. It therefore follows that these infants will pass OAE screening, which tests for an intact system of external hair cell function, but fail screening tests based on ABR measurement. During 1999, 3 of 86 newborns with confirmed hearing loss (3.5%) received a diagnosis of auditory neuropathy. (As 5 of 57 participating Colorado hospitals screened first with OAE, this 3.5% estimate of the frequency of auditory neuropathy may slightly underestimate the true occurrence rate.) Screening programs that offer initial screening with any OAE technology (including 2-stage screening programs) will fail to detect this fraction of affected newborns. In addition, as auditory neuropathy is more commonly discovered in graduates of the newborn intensive care nursery, some have suggested that ABR technology is the preferred methodology for this population.

The American Academy of Pediatrics Task Force also recommends that all infants who fail screening be recalled for adequate rescreening and follow-up, with a threshold of 95% as a standard for a successful program. During 1999, of the 46 hospitals that were screening throughout the calendar year, 9 were successful in recalling 95% or more of the infants who failed initial screening and an additional 8 hospitals were able to recall between 80% and 95% of the infants with an abnormal hospital-based screen. The largest birthing hospitals were able to achieve the highest rates of successful rescreening, perhaps secondary to their increased experience in systems management and population-based care.

Including all 60 birthing hospitals in the state, recall for reevaluation was achieved for 76% of newborns who failed their initial screening in the hospital. In earlier data from Colorado, spanning years 1992 through 1996, only 48% of those who failed the initial screening (1296 of 2709 infants) had documented follow-up testing.4 This earlier disappointing rate has been attributed to flegling data systems and reporting requirements, failure of some audiologists to report patients who returned but passed rescreening, and inadequate communication to medical professionals of the importance of timely follow-up. The current recapture rate shows clear improvement, and the true rate is likely to be higher than the 76% reported, as some infants return to a nonhospital setting for retesting. Although complete follow-up is the only logical goal for these infants, this rate of 76% compares favorably with published rates of documented follow-up for abnormal metabolic disease screening studies.20

In addition, the task force recommends that infants
who are deaf and hard of hearing be identified by 3 months of age. The Colorado median age of diagnosis of 2.1 months indicates that this goal is within reach not only for individual hospitals but also for broader hospital systems. Of the 86 affected infants who were born during 1999, 61 (71%) were 3 months of age or younger at the time of diagnosis; only 7 newborns with congenital hearing loss (as evidenced by abnormal newborn screening) had confirmatory diagnosis completed later than 6 months of age.

Federal legislation has been proposed to advance newborn hearing screening across the United States, but success has been measured only in the recent funding of federal study efforts rather than specific screening requirements. Imperfect penetration across newly participating states suggests the need for additional evaluation of a nationalized approach to newborn hearing screening. Furthermore, the continuing state-to-state variability in newborn metabolic screening efforts is an indication that broad-based national efforts to address all newborn screening efforts should remain an area of emphasis.

The successful legislation in Colorado mandating newborn hearing screening was clearly the catalyst that allowed the state to overcome a plateau in hospital participation. The dramatic increases in the number of participating hospitals and the number of confirmed hearing loss reports in infants were clearly associated with an imposed deadline (July 1, 1999) for improved penetration of hospital-based screening. Although the reality of political compromise must be considered when advocating for legislation, our experience and the recommendations of most authorities suggest that a guideline for screening every newborn is far preferable to a percentage goal. In addition, the hesitation of hospitals to participate might be better overcome by more fully addressing reimbursement issues. Data systems must, of course, support the efforts not only to screen all newborns successfully but also to ensure necessary follow-up. It is interesting that in Colorado, we found that data to measure programmatic success were more easily collected than data that would allow active intervention to ensure follow-up (eg, the name of the infant’s doctor of record, rather than the name of the mother’s doctor, rather than the name of the infant’s doctor of record).

A positive result of Colorado legislation has been the establishment of the Colorado Infant Hearing Advisory Committee. This group, in existence since 1997, has gradually influenced the program’s data collection and patient recall methods, and actively supported educational forums for physicians, audiologists, and other health professionals. The committee has drafted and recently updated guidelines for the state newborn hearing screening program, including individual guidelines for appropriate technologies, screening and follow-up, interpretation of screening results, audiologic assessment of infants, medical evaluation after hearing loss confirmation, and amplification for infants. The guidelines are expected to be available on the web site of the Colorado Department of Public Health and Environment (www.cdphe.state.co.us).

Colorado has clearly been at the forefront of national and international efforts to achieve a viable universal newborn hearing screening system. Although more than half of all 50 states have passed some form of legislation to encourage or study universal newborn hearing screening, far less than half of all US newborns are currently receiving hearing screening before hospital discharge. Colorado’s demonstration of successful population screening will undoubtedly serve to encourage others to overcome the sometimes daunting barriers to initiating universal newborn hearing screening. Support for the initiative is increasingly clear, and confirmatory reports continue to be published. As with preventing the developmental delays previously associated with congenital hypothyroidism or phenylketonuria, it is time to accept nothing less than complete population-based newborn hearing screening, thorough follow-up for infants who fail their initial testing, and timely intervention for deaf and hard of hearing newborns.

REFERENCES


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