

Newborn Hearing Screening Project, 2012-2014 on the Threshold of Effective Population-Based Universal Newborn Hearing Screening

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Abstract

Objective. Although previous studies have documented the feasibility and benefits of universal newborn hearing screening, 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: Sree Balaji medical college & hospitals voluntarily performed hearing screening before hospital discharge on all newborns from 2012 to 2014. 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. The cumulative study data as well as the results from calendar year 2012 to 2014 were reviewed for collective measures of successful screening and follow-up used otoacoustic emission testing. 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 3-year study period, 2012 to 2014, 1000 newborns were screened. A total of 150 infants who were born during the study period received a diagnosis of congenital hearing loss. In this cohort of 150 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%).

The median age of diagnosis of congenital hearing loss 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 program planners, and data systems that more accurately track and recall infants who fail initial hospital-based screening.

INTRODUCTION:

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.¹ 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.² Many physicians, however, received these new 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.³ Nevertheless, evidence in support of this aggressive universal screening Recommendation accumulated, as increasing numbers of hospitals implemented newborn hearing screening programs. With early detection and treatment of an increasing number of children with congenital hearing loss in India, comparative

developmental outcomes could be more critically assessed. In research that has subsequently been confirmed by other investigators,⁵ Yoshinaga-Itano et al⁶ 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.⁷ 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.⁶

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.^{4,15–17} This article reports the results of a screening program conducted in our hospital in the last 2 years.

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 2012-2014.

Parents 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.

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

During the study period, 150 infants were screened and 20 infants were identified as having congenital hearing loss. Of the group of infants who returned after an abnormal screening test 14 infants (71%) had bilateral congenital hearing loss subsequently confirmed, 6 had unilateral sensorineural hearing loss. Mild hearing loss (35–40 dB threshold) was present in 2, 8 had moderate hearing loss (41–70 dB), 6 had severe hearing loss (71–90 dB), and 4 had profound hearing loss (91 dB or greater). The median age of diagnosis for affected was 2.1 months. Of the 20 affected children, 10 had confirmatory diagnosis by 1 month of age, and the rest of 10 infants had confirmatory diagnosis by the fifth month of life, and the majority of affected newborns had bilateral hearing loss; fraction of 71%. Similarly, the majority of infants who had a hearing impairment (82% cumulatively) had sensorineural hearing loss.

DISCUSSION

Screening every newborn for congenital hearing loss is an undertaking of no small measure. The recruitment of diverse hospitals of varying size to participate in a statewide effort should be made feasible. 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 8 infant, 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. 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%. In addition to improving technology and experience-based training protocols implemented during the past decade, 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. 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 cochlea and external hair cells are intact but the "retrocochlear" 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.

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.

In addition, the task force recommends that infants who are deaf and hard of hearing be identified by 3 months of age. Our 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.

Our 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.

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