Children With Sensorineural Hearing Loss After Passing the Newborn Hearing Screen

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Objectives: To identify and describe the findings of children who passed their newborn hearing screen (NHS) and were subsequently found to have childhood hearing loss.

Setting: Academic tertiary care center.

Design: Retrospective medical chart review.

Methods: With approval of the institutional review board, hospital records were reviewed for children diagnosed as having hearing loss. We identified 923 children with hearing loss from 2001 to 2011. Patients who passed the NHS with subsequent hearing loss were included.

Results: Seventy-eight patients were included in our study. The suspicion of hearing loss in patients who passed the NHS was most often from parental concerns (n=28 [36%]) and failed school hearing screens (n=25 [32%]). Speech and language delay and failed primary care physician screens accounted for 17% and 12%, respectively. Configuration of the audiogram was bilateral symmetric (n=42 [54%]), bilateral asymmetric (n=16 [21%]), and unilateral (n=20 [26%]) loss. Thirty-seven patients (47%) had severe or profound hearing loss. The etiology was unknown in 42 patients (54%); the remaining was attributed to genetics (n=13 [17%]), anatomic abnormality (n=11 [14%]), acquired perinatal (n=9 [12%]), and auditory neuropathy (n=3 [4%]).

Conclusions: This is the largest study to characterize children with hearing loss who passed the NHS. In our review, parental concerns and school hearing screens were the most common method to diagnose hearing loss after passing the NHS. Families and primary care physicians may have a false sense of security when patients pass the NHS and overlook symptoms of hearing loss. This study raises the question whether further screens would identify hearing loss in children after passing the NHS.


Hearing loss is the most common birth defect and occurs in approximately 1 to 3 of 1000 infants. The incidence increases to 2 to 4 per 100 if the infant has been in the neonatal intensive care unit. The Joint Committee on Infant Hearing (JCIH) was established 30 years ago to explore the complexities of hearing loss and its effect on child development. Delay of hearing loss detection and intervention can lead to impaired cognitive development and speech and language delay. In 1972, the JCIH recommended newborn hearing screening (NHS) in high-risk infants. This criterion was further modified over the years to follow. In 1994, the JCIH recommended universal newborn hearing screening (UNHS) prior to hospital discharge for all infants. These recommendations were approved by the US Preventive Services Task Force in 2000 because of insufficient evidence favoring UNHS. The UNHS program was passed in 2001 in the state of Pennsylvania. These newborns receive their initial hearing screen prior to discharge. If they do not pass this screening, it is recommended that they will receive a follow-up examination as an outpatient within the first 30 days of life. The goal is for hearing loss to be diagnosed within the first 3 months of life and an intervention to be initiated before age 6 months.

Since the integration of the UNHS, there has been dramatic improvement of early identification of children with hearing loss. Prior to UNHS, the mean age of suspicion of hearing loss was 18.8 months, with confirmation at 26 months and hearing aids at 30 months. With the early implementation of UNHS, the age of intervention has decreased to 6 months, and in Colorado, the mean age...
for hearing aids was 5 weeks. This earlier intervention with hearing aids, bone-anchored hearing devices, or cochlear implants has decreased the amount of speech and language delay and improved cognitive development.

Physiologic measures must be used to assess newborn hearing. Either otoacoustic emissions (OAEs) or automated auditory brainstem response (ABR) testing provide noninvasive recordings of physiologic activity underlying normal auditory function. Some programs have adopted using a combination of technologies to screen infant hearing (a 2-stage screening protocol). First, an OAE test is performed. This test evaluates the peripheral auditory systems function, especially the cochlea. Low-intensity sounds are produced by the outer hair cells. When performing OAE testing, small microphones are placed in the external auditory canal, and a series of clicks are used to elicit the response. If the patient passes this test, no further intervention or follow-up is required. If they fail, repeated OAE tests are performed, often after discharge from the hospital. If the patients continue to fail, then an automated ABR test is recommended. In the ABR test, transient acoustic stimuli are generated and detected with surface electrodes that are placed near the ears. If the patients pass this test, no follow-up is required; however, if they fail, they must be followed up as an outpatient for repeated testing. Both of these screening tools have been shown to accurately diagnose moderate to profound sensorineural hearing loss (SNHL).

There have been a few studies that have evaluated patients who initially fail the UNHS and are then found to have hearing loss. Johnson et al reviewed the results of 1317 ears of patients who initially failed the OAE test and then passed the ABR test. These patients were reevaluated at 9 months, and 30 ears (21 patients) were identified as having permanent hearing loss. Seventy-seven percent of patients had mild hearing loss. Mild hearing loss may often be missed during the UNHS because the primary goal of the UNHS was to identify infants with moderate to profound hearing loss, and the equipment is tailored for this objective.

Furthermore, a recent study performed by Young et al discussed the limitations of UNHS in patients with profound hearing loss who required cochlear implantation. Two groups were analyzed: patients with implants prior to the UNHS mandate and those after the mandate. Of the 108 children in the postmandate group, 33% had passed the UNHS. They found that the average age of implantation for these patients was the same between the premandate group and patients in the postmandate group who passed the UNHS. These patients were implanted at an average age of 2.6 years, compared with 1.7 years in patients who failed the UNHS. Similarly, Wiechbold et al performed a retrospective medical chart review of 105 children with bilateral moderate to profound postnatal hearing loss and found that 23 patients (22%) initially passed their UNHS.

The literature indicates that there is a small percentage of patients who initially pass their UNHS go on to having hearing loss. In our study, we describe children who have passed both OAE and/or ABR screening tests but then were found to have SNHL.

This was a retrospective medical record review from 2001 to 2011 at the Children’s Hospital of Pittsburgh of the University of Pittsburgh Medical Center (UPMC). Approval of the institutional review board at UPMC was obtained prior to the study onset. All pediatric patients with the diagnosis of hearing loss were initially reviewed. Only patients who were reported to have undergone UNHS, which they initially passed by parent report and were then subsequently found to have hearing loss, were included in our study.

The medical records were reviewed for all eligible patients, and a database was established recording the following variables:

1. Demographics: age of diagnosis, sex, laterality
2. Results and type of the UNHS
3. Prompted further evaluation for hearing loss
4. Audiogram results
5. Neonatal risk factors: prematurity, neonatal intensive care unit stay, hypoxia, hyperbilirubinemia, and use of intravenous antibiotics
6. Other birth and medical history
7. Family medical history
8. Diagnostic test findings: computed tomography and/or magnetic resonance imaging, GJB2 and GJB6 genetic testing, Pendred testing, and other laboratory testing
9. Cause of hearing loss, as determined based on genetic, laboratory, and radiologic testing as well as history.

These data were entered into an Excel spreadsheet (Microsoft Corporation). Descriptive and inferential analysis was performed.

Of the 923 patients, 314 had undergone the UNHS and 78 patients met our inclusion criteria of passing, as reported by the parent, but were later found to have hearing loss. Of these patients, 40 (51%) were female and 38 (49%) were male. The mean age of diagnosis was 4 years and 6 months, ranging from 1 month to 10 years. Postnatal hearing loss was most commonly first identified by parental or subjective concerns in 28 patients (36%). Failed school hearing screens accounted for the second most common tool to identify this population, accounting for 25 patients (32%). Surprisingly, failed primary care physician hearing screens only accounted for 9 patients (12%), and 13 patients (17%) were identified owing to speech or language delay (Figure 1).

A majority of patients (n=42 [54%]) had bilateral symmetric SNHL. Twenty patients (26%) had unilateral hearing loss, and 16 (21%) had bilateral asymmetric loss. There was a high incidence of profound hearing loss in this cohort (n=26 [33%]), while 25 patients (32%) had mild, 16 (21%) had moderate, and 11 (14%) had severe hearing loss. The mean age of diagnosis was 6 years (range, 1 year 9 months to 10 years) for mild, 5 years 6 months (range, 1 year 2 months to 9 years 10 months) for moderate, 4 years 7 months (range, 1 year 10 months to 9 years 4 months) for severe, and 4 years 7 months (range, 1 month to 5 years 5 months) for profound hearing loss. We were able to determine the type of UNHS in a small group of patients; 12 underwent OAE testing and the other 7 had an ABR test.
The cause of hearing loss was attributed to genetics in 13 patients (17%), mainly due to the connexin mutation (n=8), but 2 patients were diagnosed as having syndromes (1 had Alport syndrome and 1 had Pendred syndrome) and 3 patients had a family history of hearing loss and declined genetic testing. A structural abnormality was identified on radiologic imaging in 11 patients (14%): 4 patients had an enlarged vestibular aqueduct, 3 had the Mondini triad, 3 had severe vestibulocochlear dysplasia, and 1 had cochlear nerve hypoplasia. An acquired perinatal cause was responsible for 9 cases (12%), due to prematurity, intensive care unit stay, ototoxic medications, or postnatal infections. Three patients (4%) had auditory neuropathy spectrum disorder. There was no identifiable cause in 42 patients (54%) (Figure 2).

**COMMENT**

With the advent of the UNHS, we are able to identify children with hearing loss at a relatively young age. Early recognition has led to early intervention and management with preferential school seating, hearing aids, and cochlear implants. This is crucial because there is a critical period for optimal language skills to develop, and with early intervention, patients have better outcomes. The literature suggests that diagnosis and intervention prior to 6 months can improve speech and language.8,10,12,14,18,19

Despite the success of the UNHS, there are some drawbacks to the screening system. It can produce a large number of false-positive results and provoke unwarranted anxiety in parents. False-positive results have been reported to be up to 30% if it is a 1-step program using only OAE testing, and decreases to less than 1% if the 2-step process is used. For both ABR and OAE testing, pathologic conditions of the outer or middle ear, motion artifact, lack of standardization, and operator-dependence are significant limitations. Patients who have this test performed while sleeping and in quiet rooms may have a more accurate test result.8,16

There is also a group of patients who have later-onset hearing loss, despite passing the UNHS.3,15,17 This population has not been very well studied; there are currently no strategies to detect hearing loss in these patients. Hence, these patients have a later diagnosis of hearing loss, leading to a later intervention and potential increased speech and language delay and cognitive and social skill impairment.12 We hypothesize that these patients can be separated into the following 4 categories: progressive or acquired hearing loss, false-negative screening result, false interpretation of screening by the operator, and miscommunication and/or misinterpretation of results given to the family.16

At our institution we use paid technicians who are trained in administering the UNHS. However, at other institutions it can be performed by a variety of individuals, such as floor nurses, volunteers, or patient care technicians, who are not experienced audiologists.14,20 The amount of training that is received by the staff is variable and may not always be adequate. This may lead to incorrect performance of the test, as well as misinterpretation of the results. If these results are not adequately documented or interpreted, then they may be relayed to the parents incorrectly. In addition, given the overwhelming experience of hospital childbirth, parents may often overlook the hearing screen results or not fully understand the results.

The UNHS can also produce false-negative results. For instance, patients with auditory neuropathy spectrum disorder may initially go undiagnosed as having hearing loss. They may have normal hearing; however, their word recognition is poorer than expected, and their acoustic reflexes, both ipsilateral and bilateral, are either absent or elevated. These children may also have normal cochlear function but a neural hearing loss. Therefore, they can have a normal OAE test result but an abnormal ABR test result.21 If an ABR test is not performed in these patients, then they may receive a diagnosis at a later date. There were 3 patients from our study with this disorder. There is a higher incidence of neonatal intensive care unit graduates with auditory neuropathy spectrum disorder.

![Figure 1. Reason for initial otolaryngology consultation for hearing loss.](image1)

![Figure 2. Cause of hearing loss.](image2)
Current JCIH recommendations are that neonatal intensive care unit babies admitted for more than 5 days must be screened with the ABR test only, since neural hearing loss may be missed with OAE screen.22

Patients with an enlarged vestibular aqueduct are reported to have hearing impairments that can be fluctuant and progressive.23 It can occur in a stepwise onset or suddenly, triggered by the Valsalva maneuver, minor head trauma, increased barometric pressure (scuba diving), jogging, or even common colds.24 Onset of SNHL can range from birth to adolescence but is mostly noted in the first few years of life.25,26 Seven patients in our cohort had an enlarged vestibular aqueduct, with 3 of these 7 having a Mondini triad (1 of these patients had Pendred syndrome). The onset of hearing loss for these patients ranged from 2 years 6 months to 7 years 7 months, and the severity ranged from mild to profound. There was no noted inciting event for hearing loss in these 8 patients in our group.

Furthermore, 9 patients (12%) in our cohort had an acquired cause—prematurity, intensive care unit stay, ototoxic medications, and postnatal infections. Their ototoxic effects may have started to take place after the UNHS was already performed. Unfortunately, we do not have the exact timing of each hearing screen to evaluate if the patient had received antibiotics or had infections prior to the administration of the UNHS. In these cases, patients may have been born with normal hearing at birth and at the time of the test.

Also, the incidence of cytomegalovirus infection (CMV) is 0.2% to 2.2% for live newborns,27 approximately 30% to 50% of which have clinically apparent SNHL at birth.28,29 Onset of hearing loss in patients with CMV can be immediate or delayed with variable severity.27,28 Fowler et al29 looked at 388 children with congenital CMV and found that only 5.2% of patients had hearing loss detected at birth. They found that late-onset hearing loss occurred throughout the first 6 years of life in approximately 10.2%. Some of our patients may have had CMV with a later onset of hearing loss; however, we do not have these laboratory results to confirm this.

Both the ABR and OAE screens have been geared at detecting moderate to profound hearing loss.6,39 Often, patients with mild hearing loss at birth will not have hearing loss detected. Johnson et al40 found that of those patients who passed the UNHS and were subsequently found to have hearing loss later in childhood, 23 (77%) had a mild loss.15 However, in our population, only 25 patients (32%) had mild hearing loss. Thirty-seven patients (47%) had severe to profound hearing loss. Some of these patients may have been missed at the initial testing, while for others, the hearing loss may have progressed over time.

Because the UNHS has been seen as a highly effective screening tool, patients with hearing loss despite passing may be overlooked. Parents, pediatricians, and family physicians may have a false sense of security from a normal hearing screen result and may overlook signs of hearing loss. This may account for why only 9 of the referrals (12%) were after failed screenings from the primary care physician’s office. The rate of referrals from parental concerns was 36%, most likely because parents often will compare the development of siblings and other children in their environment on a more frequent basis, and they are directly affected by the consequences of the hearing loss. Weichbold et al41 also found that parental concern most often prompted hearing assessment, with 57% in their patient population with postnatal hearing loss after initially passing the UNHS. Failed school hearing screens were also very useful in determining patients with hearing loss, accounting for 25 of the referrals (32%) from our cohort. However most of these screenings were performed in preschool, kindergarten, or the first grade. In our patient population, the mean age of diagnosis of severe to profound hearing loss was 4 years 7 months, ranging from 1 month to 9 years 4 months. It was slightly higher for moderate (5 years 6 months) and mild (6 years). If their actual onset of hearing loss was early, these children are not benefiting from early intervention, which is one of the major goals set by the JCIH.

The use of additional screens after the newborn period remains to be investigated. Costs and the utility of screening results have to be considered before universal implementation of such programs. Currently, most schools have a screening system in the beginning of each year starting in preschool. However, this would mean that children could potentially go undiagnosed until they are 4 years of age. An earlier screen at the 3-month wellness visit is currently being studied in our institution. We hope that this will help to identify the remainder of the hearing loss population and provide hearing supplementation for them. It may also serve to identify infants who were lost to follow-up. Current Centers for Disease Control and Prevention reports demonstrate that approximately 45% of infants who did not pass newborn screens do not receive the appropriate follow-up.30 We also believe that it is important to standardize the OAE and ABR equipment used for the screening infants across the nation. Along with standardizing the equipment, we believe that it is important to also have a regimented training program for anyone who is to administer the test so that they understand not only how to perform the test but how to interpret the results and discuss the results with the family.

Our study had some limitations as a retrospective medical review. We were not able to confirm the UNHS results; most of these data came from parental report to the audiologist and pediatric otolaryngologist and were recorded in the patient medical chart. We were only able to identify if the patient underwent either an OAE or ABR screen in 19 of the 78 patients. These data would have been useful to further analyze if there was more false-negative results noted with either testing modality. This low number also raises the possibility that the child actually did not pass the screen and the family had misinterpreted the results. Regardless, the delayed presentation to our tertiary medical center was based on the family’s and primary care physician’s understanding that the child had passed the newborn screen. In addition, not all patients had laboratory, genetic, or radiographic testing performed. Only 39 patients underwent genetic testing, and 67 patients had a computed tomographic scan, while 50 received magnetic resonance imaging. If we were
able to obtain these data uniformly in each patient, we may have been able find a cause in more of our cohort. Also, because these patients were referred to our tertiary care center from several hospitals, we were unable to get an accurate percentage of patients who passed their UNHS and were later diagnosed as having hearing loss, since we do not have the data for all the patients who had undergone the screening and their results.

In conclusion, this is the largest study to characterize children with hearing loss who initially passed the UNHS. Although the UNHS has been instrumental in diagnosing hearing loss in newborns and allowing for earlier intervention, it has its limitations. This study highlights the importance of recognizing the possibility that children who passed the UNHS may have hearing loss. Parents and medical care providers should recommend further audiologic testing in children with speech and language delay regardless of passing the UNHS. Further studies to investigate the use of additional hearing screens are required.

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REFERENCES


