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Newborn Hearing Screening in 2010

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Abstract

Purpose of Review—The objectives of this review are to provide the reader with a current and concise review of the data and trends in universal newborn hearing screening(UNHS). Within a relatively short period of time, the concept of screening all infants for hearing loss at the time of birth has evolved from a nascent process to a truly universal system in most developed countries. As a result, the focus and challenges of UNHS have shifted to topics of developing ever more efficient and cost-effective approaches and potentially melding physiologic hearing screenings with ancillary screening techniques.

Recent Findings—Enhancement of the UNHS process is likely to be accomplished by implementation of novel tools such as wideband reflectance technologies and intelligent incorporation of screening for common genetic and viral causes of congenital hearing loss.

Summary—With such a rapidly evolving process, it will be critical for clinicians to understand the benefits and limitations of various newborn hearing screening methodologies in order to determine the most appropriate management of children referred from their UNHS. This will entail a working knowledge of emerging audiologic tools as well as infectious and genetic etiologies of pediatric hearing loss.

Keywords

Newborn Hearing Screening; Hearing Loss; Congenital; Cytomegalvirus; Wideband Reflectance; Auditory Neuropathy; Auditory Dyssynchrony

Introduction and Issues Related to Newborn Hearing Screening

Less than one decade ago, the concept of implementing universal newborn hearing screening (UNHS) protocols was an active topic of debate. The merits of physiologically testing every newborn were contested given the potential for false positive referrals, the stressors imposed on parents, audiologists, early interventionists and physicians, and the costs burdened upon birthing facilities and an already strained medical economic system. In favor of newborn hearing screening, proponents argued that congenital handicapping hearing loss affects as many as 2 per 1000live births (http://www.nidcd.nih.gov/health/statistics/quick.htm) making it the most common neurologic birth defect in the United States. The negative impact of missing an infant's congenital hearing loss on that child's communicational, social and

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emotional development are well-documented. Earlier education and cost-to-society studies demonstrated that the late identification of a child with permanent severe-to-profound hearing loss costs the educational system anywhere from $38,000 – $240,000 per child over the span of a Kindergarten through 12th grade education in terms of special education interventions [1, 2].

However, in just a short span of time, those issues have mostly been cast by the wayside as UNHS protocols and systems are implemented worldwide. Data from local programs as well as Centers for Disease Control (CDC) Early Hearing Detection and Intervention (EHDI) programs indicate that in many areas, the number of infants identified with congenital hearing loss has doubled when compared to pre-UNHS time points. With this leap forward, the focuses have shifted to finding the optimal times, personnel, equipment and data management systems required to execute UNHS efficiently and effectively. And in similar fashion, attention has now been directed at the challenge of reducing lost-to-follow up issues when infants are referred from their newborn hearing screening and fail to follow through with the recommended diagnostic audiologic testing(as well as management). In many regions, this lost-to-follow up rate approaches 40% of infants not passing their newborn screening.

In very short order, many arguing points regarding UNHS have been either surpassed, attained a point of consensus agreement, or in some instances, continue to be debated without any possibility of reaching consensus. Points such as the feasibility of screening, the optimal technology for screening, the personnel that should be performing the screening, etc, have all been well-reviewed in the literature. Accordingly, this Current Opinion section will attempt to address more emerging concepts and technologies that may become pertinent to UNHS while still acknowledging and addressing the pragmatic considerations for universally screening infants in the bleak medical economic environment. With very cautious optimism, it is tenable to speculate that emerging (and cheaper) technologies that exploit our growing knowledge base concerning congenital hearing loss etiologies, might actually allow NHS to function as a major cost-saving intervention in our society and medical system.

Technologies Relevant to Infant Hearing Screening

Most birthing facilities in the US, the European Union, Australia and developed Asian countries have evolved towards implementation of Otoacoustic Emission (OAE) and/or Automated Auditory Brainstem Response (AABR) testing as the methodology of choice for performing newborn hearing screening [3–8]. Advantages and disadvantages to each technique create proponents for use of each screening methodology and are reflected in the table below (Table 1).

Pragmatic considerations include the personnel who can perform the hearing screenings in the newborn nurseries or other pediatric units. In some instances, trained Audiologists (or trained Audiology aids) perform the screenings; an option that offers the greatest level of expertise and likely provides the most effective screening. In contrast, however, many birthing facilities do not have the luxury of utilizing Audiology personnel and instead use trained volunteers, patient care assistants or clerical staff on the units. Many centers also add the responsibility of performing the NHS onto nursing staff in the nurseries. Any meaningful screening technology requires a high degree of ease-of-use and sufficient sensitivity and specificity such that a screener with minimal training and experience can efficiently and accurately perform the NHS. Both OAE and AABR techniques meet these core requirements and accordingly have become the widely accepted practices around the world [7, 9–15].

Auditory Neuropathy Dyssynchrony Disorders

A common discussion in this topic area of UNHS is the challenge of identifying and diagnosing infants with Auditory Neuropathy Dyssynchrony Disorders (ANDD) [5, 16–19]. The typical child with ANDD may display robust OAEs but have either an absent or markedly dysmorphic AABR response in combination with varying degrees of hearing loss (as demonstrated by behavioral threshold testing). In such instances, an infant screened using OAE techniques would pass their UNHS and potentially be missed until significantly later ages. Most experts in this field concur that a full audiologic battery (that includes OAE and ABR testing) provides the greatest sensitivity and specificity for ANDD. This is typically beyond the scope of the newborn hearing screening protocols but certainly poses a strong argument for incorporating “two-step” or “re-screening” algorithms into any UNHS protocol. In many birthing centers, an established practice is to perform an initial screen using the most cost-effective technique (typically transient evoked OAE or distortion product OAE). If an infant does not pass the initial screen, a common protocol is to then switch to AABR in order to perform a re-screen. This type of NHS protocol offers the advantages of evaluating the auditory system in two different manners and potentially increasing the sensitivity for an ANDD. In some facilities, an OAE screen is performed twice (at time points that are separated by several hours if possible prior to discharge) with the rationale that transient noise, activity (crying) or fluid factors had resulted in a false positive at the first screen. Two-step screen/rescreen protocols have been demonstrated to help reduce the false positive referral rate from NHS but obviously impose twice the burden to the birthing facility [20].

The Future of Universal Newborn Hearing Screening

Technologic advances in simple and accurate audiometric evaluation of the newborn clearly present some opportunities for improving the sensitivity and specificity of UNHS. Perhaps a prime example is demonstrated by the development and application of wideband reflectance(WBR) as a physiologic and clinically useful test of middle ear function in infants. By developing simple broad band reflectance devices for use in nurseries and other infant care units, several investigators suggest that WBR can help reduce the false positive referral rate such that the subsequent diagnostic audiology burden on audiologists and the healthcare system is significantly reduced by eliminating many of the infants where persistent amniotic fluid or a routine middle ear effusion, or even vernix in the ear canal, cause Otoacoustic Emission or Auditory Brainstem Response tests to yield false referrals on these children [21–35].

Cytomegalovirus and Genetic Screening in Congenital Hearing Loss

Our understanding of the role of congenital viral infection and hearing loss, and specifically congenital Cytomegalovirus (CMV) infection, has improved dramatically over the past few years and reports cite as many as 1% of all infants born in the US are infected with CMV [36–44]. Sensorineural hearing loss (SNHL) represents one of the most common sequelae of congenital CMV infection in infants and another large group of children with so-called “asymptomatic CMV infection” may present with hearing loss alone and no other CMV-related manifestations (e.g. no hepatosplenomegaly, petichiae, retinitis, microcephaly, brain calcifications, etc)[36, 40, 45]. Making this topic even more clinically compelling are newer data now demonstrating that treatment of children with congenital symptomatic CMV infection (with intravenous ganciclovir)can stabilize or even rescue hearing if the CMV infection is diagnosed and treated early [36–38, 46]. As a result, clinicians are now presented with the challenge of quickly identifying infants with congenital CMV infection (and CMV-

related hearing loss)in order to allow delivery of medical therapies that might actually stabilize or improve hearing for these infants.

One of the best windows of opportunity to screen for congenital CMV infection would be at the newborn period. Preliminary studies have already documented the feasibility of using blood spots from a Guthrie card as a DNA sample for polymerase chain reactions(PCRs) that would allow amplification of viral DNA sequences which in turn would indicate congenital CMV infection. Alternatively, simple and non-invasive sampling of infant saliva is another excellent source for culturing or sampling of virus since CMV is typically concentrated in salivary secretions [47, 48].

Such a scheme obviously suggests a “hybrid” newborn hearing screening process in which physiologic hearing testing (OAE, AABR, wideband reflectance) is coupled with clinical laboratory-type screening methodologies to enhance the sensitivity and specificity of the newborn “hearing” screening. While such laboratory testing has traditionally been delegated to the diagnostic workup phase of an infant with congenital hearing loss, it is worthwhile considering the benefits of exploiting new diagnostic technologies and new knowledge of virally-mediated hearing loss and its treatment, to enhance the screening/diagnostic process and potentially reach a successful treatment phase in a shorter time frame. Taking into consideration the substantial lost-to-follow up challenges faced by most newborn hearing screening programs, it is even more compelling to consider performing as much screening, diagnosis and counseling or intervention as possible while the infant and family is engaged in the medical system. Further data are needed to determine the frequency of CMV-related hearing loss in infants as part of the calculation of whether CMV screening merits inclusion in a newborn hearing screening algorithm. This exact data is currently being collected in a large scale, multicenter study funded by the National Institutes of Health (NIH), National Institute on Deafness and Other Communication Disorders (NIDCD) that seeks to enroll 100,000 infants that will be screened for congenital CMV infection (http://www.nidcd.nih.gov/health/inside/spr06/pg3.htm). This information will become increasingly pertinent as more efficient and cost-effective methodologies for CMV screening are developed. A later potential benefit of congenital CMV screening would also be the identification of those children who are at elevated risk for later onset hearing loss due to CMV infection. By age 6 years, 6 per 1000 children will display a permanent hearing loss. At the present time, our knowledge of who is at higher risk for later onset hearing loss is minimal and identification of those children is difficult or often delayed.

It is completely tenable to foresee further expanding this hybrid screening concept to include molecular screening methodologies that could analogously utilize either Guthrie card bloodspots or simple buccal brushings that provide adequate DNA samples for mutation screening. With the ever increasing availability and utilization of molecular genetic testing for deafness causing mutations, it is plausible to envision a molecular screening process that utilizes rapid PCR assays to detect common mutations (e.g. 35delG of GJB2). With reports of GJB2 mutations being responsible for anywhere from24–40% of pediatric patients with a confirmed SNHL [49–51], it would be reasonable to suggest incorporating a limited and rapid molecular screening for hearing loss mutations in an innovative UNHS protocol.

An inescapable aspect of such molecular or CMV screening, and indeed, all NHS methodologies, is the cost factor. With most births being covered by a capitated reimbursement system and no additional reimbursement being provided for NHS, minimizing costs to the birthing facility is essential [20, 52–58]. The cost of molecular diagnostics have the potential to gradually decrease as robotic PCR and automated sequencing systems are implemented that allow higher throughput of samples with less manpower utilization and some economies of scale. However, interpretation of molecular

diagnostics remain a key feature that necessarily involves greater manpower time, effort and expertise. In the case of CMV screening at birth, work on refining different assays such as PCR of viral sequences, rapid microcultures, or routine viral cultures, will determine how cost-effective viral screenings may become.

The Political Landscape in UNHS

As a final consideration in this discussion, it is requisite to discuss the common political landscape (particularly in the US, but perhaps globally) with regards to governmental funding, oversight and support for UNHS programs. Due to the inherently medical nature of NHS in the neonatal period, governmental oversight of NHS programs typically fall under the domain of the departments or ministries of health. Costs related to the newborn screening, the data management from screenings, as well as any early interventions triggered by the identification of hearing loss in infants and children, similarly fall within the budgetary scope of a health branch of government. However, some of the most significant outcome measures that reflect the effectiveness of early identification (UNHS) and intervention for congenital hearing loss, are the educational readiness and performance when those children reach Kindergarten and elementary school levels. As a consequence, the early efforts and expenses incurred by a department of health, may not show a proximate “return on investment” that would politically provide the justification for continued support of UNHS programs. Such “disconnects” between one agency’s upfront investment and a different agency ultimately acting as the recipient of those benefits can create inherent political challenges in garnering long term support for UNHS in some circumstances. It remains the role of higher levels of government to perceive the long term concerted efforts of different branches of government in achieving the best possible outcomes for children. It is also the responsibility for medical, early intervention and education professionals to advocate for children with congenital disabilities such as hearing loss and to make the “big picture” obvious for those officials within compartmentalized branches of government.

Conclusion

As our knowledge base and understanding of the etiologies of congenital hearing loss continue to broaden, it will be essential for those advances to be translated into enhanced newborn hearing screening methodologies. Incorporation of simple, non-invasive, sensitive and specific audiologic tools will similarly be requisite in order to meet the challenges of universally screening all infants while containing/reducing costs in difficult medical economic times. Given the frequency of genetic and viral etiologies in congenital hearing loss, it is only logical to begin considering screening approaches for these etiologies in UNHS protocols. As treatments for viral and genetic hearing losses begin to emerge from the benchtop, the relevance of screening for those conditions will only continue to increase. A concomitant burden is then placed on clinicians to keep up to date on these rapidly evolving newborn hearing screening methodologies so that the information can be utilized in proper fashion and subsequent diagnostic and therapeutic decisions made expeditiously. Finally, it is very pertinent to remain focused on the ultimate objective of identifying infants with hearing loss as early as possible in order to implement early interventions and allow for better hearing, language and global outcomes for these children.

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Table 1

Comparison of OAE and AABR Newborn Hearing Screening Technologies

	Otoacoustic Emissions Testing	**Automated Auditory Brainstem Response Testing**
Advantages	Simple testing technique (relatively minimal training needed to perform OAE testing); Cheaper than AABR screening; Fast	Superior evaluation of the auditory system (vs assessment of outer hair cell function alone as in OAE); Likely provides better detection of infants with auditory neuropathy;
Disadvantages	Limited assessment of the auditory system; Impacted by middle ear fluid issues; Potentially impacted by vernix or wax in the ear canal; Optimal to perform in a quiet environment;	Requires more operator knowledge than OAE testing; Potential for electrical and noise artifact yielding poor screening; Requires sleeping or quiet infant; Optimal to perform in a quiet environment; Requires longer times than OAE screening; Typically more costly than OAE screening;