NEWBORN HEARING SCREENING IN SICILY: LESSON LEARNED

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ABSTRACT

The objective of this work carried on by University of Palermo, was to evaluate the prevalence of sensorineural hearing loss (SNHL) from 2008 to 2012 through a newborn hearing screening programme, focusing on 3863 newborns with and without risk indicators. The authors performed a global audiological assessment through otoacoustic emission (TEOAE), tympanometry and auditory brainstem responses (ABR) studying the main factors reported by Joint Committee on Infant Hearing (2007) and using the UNHS (Universal newborn hearing screening). The principal lesson learned after years of involvement in developing and managing newborn hearing screening programmes is to approach this problem with a healthy dose of realism, and to manage existing resources wisely.

***Key words**: Screening neonatale, neonati a rischio, sordità neurosensoriale, ABR. TEOAE.*

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Introduction

The importance of early detection of hearing loss followed by appropriate intervention cannot be overstated. There is no doubt that the goal should be to identify every infant with hearing loss and to intervene appropriately at a very early age[1-5]. With the advent of cochlear implants for children over 25 years ago, we have had the technology to address the entire range of neonatal hearing loss[6,7]. While this is an achievable goal in relatively wealthier countries with well-distributed resources, it is not necessarily the case when resources are limited. The question is how to best utilize existing, limited resources, and maximize those for the benefit of early treatment of hearing loss in newborn, infants and young children. We will attempt to frame this important within our experience with neonatal hearing loss and its management. Initial effort in the use of the auditory brainstem response in the identification and the characterization of hearing loss focused on premature newborns, specific conditions such as perinatal asphyxia and the incidence of hearing loss in high-risk and intensive care nursery infants[8]. Thus, the initial early hearing loss detection programmes focused on newborns with one or more risk indicators known to be associated with hearing loss. These risk indicators were revised periodically and new risk indicators were added while others were downgraded or deleted[9,10]. This trend is well-represented by reviewing risk indicators listed by the Joint Commission of Infant Hearing (JCIH) over the years. Reports on the incidence of hearing loss in initial newborn hearing screening programmes were somewhat variable as was the case in a 1985 publication comparing five Canadian newborn hearing screening programmes. Specifically in this programme the total incidence of hearing loss resulted 6.8%, but the incidence of bilateral moderate to profound hearing loss was 2.9%[11].

Materials and methods

The Audiology Department, University of Palermo, began a newborn hearing screening programme from 2008 to 2012, focusing on newborns with and without risk indicators. We performed a global audiological assessment (through TEOAE, tympanometry and ABR) studying the main factors reported by Joint Committee on Infant Hearing (2007) and using the UNHS (Universal newborn hearing screening). A review of our programme involved 3863 newborns, of these 815 presented one or more risk indicators for significant hearing loss, 3048 not have risk factors. Of 815 babies with risk factor, 56 were diagnosed with bilateral hearing loss and 46 with conductive hearing loss while of 3048 babies without risk factor, 5 were diagnosed with bilateral hearing loss and 40 with conductive hearing loss.

Discussion and conclusions

Universal newborn hearing screening (UNHS) has now been the norm for over a decade in the great majority of birthing facilities in the North America, Great Britain, Europe and Australia.

The strategy of UNHS (Universal newborn hearing screening) is to screen all newborns shortly after birth, not just those with risk indicators. The goal is to avoid delaying the diagnosis of hearing loss in an infant who may not have any known or recognized risk indicators, and who would not be included in a high-risk hearing screening programme[10,12,13]. Some authors suggested that as many as 50% of newborns with loss were being missed if only high-risk screening was being carried out. Therefore, the impetus for transitioning to UNHS from high-risk screening became stronger. With the advent of UNHS it was thought that it may no longer be necessary to identify risk indicators, however risk indicators continue to represent a critical component in the understanding of the pathophysiology of congenital hearing loss[14].

At the University of Palermo we initiated UNHS in 2008, using auditory brainstem response (ABR). However, along with successfully evaluating all newborns prior to discharge from the hospital (> 99% inclusion rate), we continued to identify and document risk indicators. Our prevalence rate resulted superimposable with the principal UNHS programme of the others country.

Hearing loss, either conductive and sensorineural, in infancy is one of the most common defects; it is well-documented that the children who are identified through universal newborn hearing screening, and diagnosed at an early age, have better language outcomes at school age than those who are identified later in life[7,8,15-17]. It is of note that the initiation of universal newborn hearing screening programme coincided with the introduction of TEOAE and ABR[2].

This allowed a technician to complete the testing as opposed to an audiologist who was responsible for ABR hearing screening when operating the high-risk screening programme. The protocol for our UNHS programme is as follows: infants are tested with ABR between 10 and 20 days of life. If the infant passes the initial screening bilaterally and has no risk indicators for delayed-onset hearing loss, no further planned testing is carried out. Those infants who refer on the initial ABR screening test receive an additional audiological evaluation while in house. Those who pass the screening test are scheduled to return in six months for a behavioral audiologic evaluation to confirm normal hearing. Those who do not pass return within two to three weeks for a diagnostic auditory brain stem evaluation, as well as behavioral testing, otoacoustic emission evaluation and tympanometry, if possible. Those presenting with one or more indicators for late-onset hearing loss are scheduled for evaluation in three to six months even if they pass the initial screening. During our first 4 years of UNHS, 3863 infants were screened as previously described. Of those, 815 had at least one for hearing loss. Of the 3048 with no identified risk indicators, 45 were diagnosed ultimately with hearing loss, among these 40 were diagnosed with conductive hearing loss with objective tinnitus in three cases (it can result from the changes in the surface tension of mucus in the Eustachian tube)[18] and 5 with sensorineural hearing loss. The statistics of the at risk population numbering 815 were quite different: 102 infants were diagnosed with hearing loss. From our total of 147 infants diagnosed with permanent hearing loss from the UNHS programme, 64% had binaural hearing loss and 36% had unilateral hearing losses[19]. The majority of babies at risk were diagnosed with hearing losses within our universal newborn hearing loss programme were sensorineural. It is of note that within the at risk population there were 46 infants diagnosed with permanent, conductive hearing loss[20-23] (fig. 1).

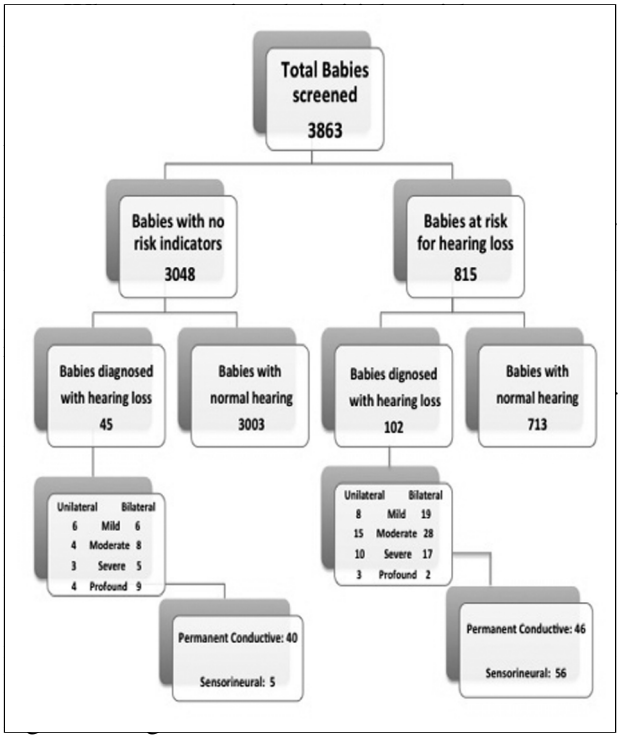

Figure 1: Outcome of universal hearing screening.

So what lessons have learned? There is no doubt that UNHS is superior to risk-indicator-based screening by identifying all hearing loss present at birth. It is clear that a significant majority of neonatal hearing loss is associated with one or more risk indicators. UNHS is more labour-intensive requiring more resources to just accomplish the initial phase, and to make a notable difference requires substantial resources for follow-up and management. Realistically, when resources are limited, they need to be allocated between the initial screening effort and the management effort. In such situations, it may be more effective to initiate a risk-based newborn hearing screening programme, with sufficient resources allocated to managing diagnosed hearing loss, and combine that with an effort to educate primary care physicians regarding hearing loss in infancy, with the hope that they would make appropriate referrals. In this way, the newborn hearing screening programme would not just be a massive effort to carry out primary screening without the resources to treat.

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