Newborn Hearing Screening: A Regional example for National Care

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Abstract
Congenital Permanent Childhood Hearing Impairment (PCHI) is known to have a negative effect on language acquisition, cognitive development and social integration. Since 2000 our department has implemented a UNHS program in the West of Ireland. We describe our experience and detail our results to date. All neonates born from October 2000 to November 2007 were screened using a 2-stage protocol. Transient evoked otoacostic emissions (TEOAEs) were used to screen all neonates, followed by automated auditory brainstem response (AABR) in those who did not pass TEOAE, and all neonates at audiological risk. 26,281 babies were born over the eight year period. 25,742 underwent the screening process, achieving a coverage rate of 98%. The prevalence of PCHI in the population tested was 1.21/1000 live births (31/25,731). Our results show that a hospital based 2-stage UNHS protocol using TEOAEs and AABR is accurate, feasible and effective.

Introduction
Congenital Permanent Childhood Hearing Impairment (PCHI) is more prevalent than many birth defects for which there is mandatory screening [1,2], affecting at least 1 baby in 1000 [3,4]. In the Republic of Ireland (RoI), PCHI occurs in approximately 1 in 813 children before 5 years of age [5]. Approximately 84 children are born in the RoI with a permanent hearing impairment per year [6]. It has been well established that PCHI has a deleterious effect on language and cognitive development, educational attainment and quality of life, as hearing is essential for the normal development of speech [7-11]. Hence, early diagnosis and intervention by 6 months of age is imperative to avoid these consequences. The lack of neonatal screening can lead to a delayed diagnosis of hearing impairment [12-15]. In some countries, only babies deemed to be at high risk of developing PCHI are screened. While other countries (eg USA, UK, Austria) have embraced and developed UNHS programs in order to achieve early detection and intervention [22-27]. The European Consensus on Neonatal Hearing states that neonatal testing in maternity hospital is more effective and less expensive than behavioral testing at 7 to 9 months of age [21]. The international experience regarding UNHS shows that these programs once up and running are cost effective, accurate, family friendly and easy to maintain. The most common methods used in UNHS programs are automated otoacoustic emissions (OAEs) and automated auditory brainstem response (AABR) [29].

In 1978 Kemp described OAEs as a means to determine cochlear outer hair cell function [33]. OAEs are a by-product of the biochemical motility of the outer hair cells. They can be recorded with a small microphone placed in the ear canal and confirm the integrity of the cochlea. Two types of evoked OAE are in use in UNHS, the transient evoked otoacoustic emissions (TEOAEs) and distortion product otoacoustic emissions (DPOAEs). AABR is frequently used for newborn hearing screening as it provides accurate information in a fairly short space of time. It provides an electrophysiological measure of the auditory pathway along the auditory nerve. To administer the AABR test, electrodes are placed on the infantâ s forehead to record the brain activity of the auditory brainstem in response to sound. The infantâ s ears are covered with earphones that emit a series of soft clicks. The AABR system compares the infantâ s response against a normal template and provides a pass or refers report. There is no nation-wide UNHS program in the RoI. The current screening system is Health Visitor Distraction Testing (HVDT) which is performed in older children (minimum age 7-9 months). The Departments of Otolaryngology and Audiology, Galway University Hospital have implemented a UNHS program in two hospitals in West of Ireland since 2000, screening 25,742 babies in total. The two hospitals involved are: University College Hospital Galway and Castlebar General Hospital, Co Mayo. Our protocol consists of a 2-stage process testing TEOAEs in all neonates and AABR in those that do not pass. Neonates at audiological risk routinely undergo TEOAEs and AABR.

Methods
The screened population included all neonates including those at audiological risk born between October 2000 and November 2007. Screening was performed in the maternity ward on babies within 48 hours of birth by audiologists (Monday to Friday). Any baby discharged prior to screening or born at the weekend was referred directly to the audiology outpatient department. Where possible all newborns were screened at the bedside in the presence of the mother. Verbal explanation of the screening process was given to the mothers. Additionally, a leaflet describing the screening was also available. Informed verbal consent was received from all mothers prior to screening and this is viewed as sufficient from a risks management point of view. A 2-stage UNHS protocol was used for well babies and babies at audiological risks (Figure 1).

At the first stage all well babies were screened in both ears using TEOAEs (ECHOCHECK ILO88 OAE system, Otodynamics Ltd). Newborns who failed the initial test were screened again using TEOAEs within 2 weeks. Outpatient screening by one month of age was also performed on all newborns discharged without screening. In stage two, if the baby fails the 2 step TEOAEs (hospital and outpatient TEOAEs), he/she undergoes AABR (ALGO 3i Newborn hearing screener, Natus medical) within 6 weeks. All babies at audiological risks in the neonatal intensive care unit were screened with both TEOAEs and AABR irrespectively of whether they pass TEOAEs. All cases who failed AABR were referred for full diagnostic ABR (Bio-Logic Navigator Pro, Natus) by the audiological department at 3 months of age. Cases with unclear results are referred to our Department of Otolaryngology for further diagnostic assessment. Sensitivity and specificity were based on the observed true positive (TP), true negative (TN), false positive (FP) and false negative (FN) cases. Sensitivity was calculated as TP/ (TP+FN) and specificity as TN/ (FP+TN).

Figure 1: UNHS protocol

Results
There were a total of 26,281 live births, of which 25,742 were screened (98% coverage). 14,653 well babies were screened in the maternity ward, 8,889 well babies had outpatient screening and 2,200 babies at audiological risk were screened in the neonatal intensive care unit (NICU). 358 (1.36%) babies were not brought to outpatients for screening and 41 (0.16%) were not screened because their parents refused. Ninety-two (0.36%) babies failed the 2 stage UNHS using TEOAEs and AABR. Of these, 50 passed diagnostic ABR, five did not attend follow up, five were relocated to other hospitals and one died. Thirty-one failed diagnostic ABR (0.12% of the total screened population had congenital hearing loss). Therefore, the prevalence of PCHI was 1.2 per 1,000 live births (31/25,731). Bilateral PCHI 40db relative to hearing threshold was found in 19 newborn (0.07%) and the remaining 12 neonates were found to have unilateral SNHL. Both groups were referred for further Otolaryngological assessment. All the 19 babies with bilateral PCHI were fitted with hearing aids. All babies were diagnosed by 3 months of age and received intervention by the age of 6 months. Our UNHS 2-stage protocol showed a 100% (95%C.I.: 86.273-100) sensitivity and 99.8% (95%C.I.: 99.7414-99.8541) specificity in detecting congenital permanent childhood hearing loss.

Figure 2: UNHS Working Group â Program of Action for Children Recommendations

Discussion
The Joint Committee on Infant Hearing (JCIH) recommended UNHS and provided guidelines in 1994, 2000 and 2007 [28-30]. The recommended standard is that hearing loss in newborns should be detected by 3 months of age and intervention implemented by 6 months of age. Also, the European Consensus Development Conference on Neonatal Hearing Screening recommended neonatal hearing screening to be the first part of a program of habituation of hearing impaired children [21]. Currently, the implementation of UNHS programs is becoming more frequent in many developed countries. It has been implemented in USA (over 38 states) [22, 23], England [24] and various European countries [25-27]. Currently, in Ireland it is not widely practiced.

HVDT at 9 months of age is the current nation-wide screening system, and was introduced in the 1950s. The average age of identification using the HVDT screening program in Ireland is approximately 30 months [5]. This results in delay in intervention which can be for many months to years. HVDT has been shown to be variable in efficacy and unable to detect congenital hearing impairment prior to 6 months of age [16,5]. Owing to staff shortages it is not easy to achieve the 7 â 10 month time scale of screening using the

health visitor distraction testing. Children who fail are generally retested 1 month after the first test. If they failed again they are referred to the community audiologists for further assessment and diagnosis. The report by Davis et al in 1997 showed that there is a poor sensitivity and relatively poor specificity for the HVDT, with relatively low yield. Median age of identification via the HVDT was found to vary from 12 to 20 months [17]. Recent published work from the South of Ireland showed that the average age of identifying hearing loss is 11- 18 months of age and the median age of fitting hearing aid is 20-49 months. This clearly demonstrates the failure of achieving early diagnosis of PCHI5.

Regarding cost implications, in 1998 Stevens et al published a survey detailing the cost implications of hearing screening. They found that the cost of UNHS was considerably less than that of the HVDT and their results suggested that the most cost effective approach to early identification of PCHL is the UNHS [9]. Other studies reported similar findings [17]. The cost of educating a child in a special school is presumably higher than for a child in a mainstream school in Ireland. Early detection and treatment of hearing loss in children will enhance the opportunity for these children to receive formal education, acquire sufficient skills to achieve vocational prospects equivalent to that of their hearing peers. Without early detection the societal cost required to provide support for these children in the long term will be high [18-21]. In 2004, the UNHS Working Group â Program of Action for Children made a recommendation that UNHS should be introduced in Ireland as a matter of urgency and outlined a critical pathway to follow (Figure 2) [6].

Our UNHS program meets the internationally set criteria and has results similar to previously published studies [19]. We achieved a screening population capture rate of 98% which exceeded the standard UNHS goal of 95% minimum capture rate as stated in 2000 by JCIH. Also, we noted that we had a false positive rate of 0.5% which is in keeping with the recommended goal of a false positive rate of less than 3%. The referral rate for audiological and medical evaluation after the screening process (during both birth admission and outpatient screening) was 0.36% which is very low. This achieves the recommended standard of <4% [20]. Our findings in the West of Ireland strongly justify the use of a 2 stage UNHS protocol. The most important aspect of a successful hearing screening program is follow up. Hence, we contacted all babies that did not attend and forwarded their names to the community audiologist, those that relocated to another area in Ireland were sent to the nearest audiological service. In RoI with increasing healthcare costs, there is a growing trend towards early discharge of patients within 24hours of delivery. It has been a challenge to maintain timely diagnostic evaluations due to inadequate audiological resources.

Availability of necessary resources is imperative in order to run and support an effective program. Tracking of patients and keeping of records is challenging especially now that Ireland is experiencing increase number of immigrants and asylum seekers. Sometimes communication can be a source of problem when dealing with this group because of language barrier. Follow up of migratory groups is a challenge. In order to ensure that early detection and effective intervention are made on all newborns with hearing impairment, UNHS should be performed. UNHS is becoming standard of care in many developed countries. Our current 2 stage UNHS protocol using TEOAEs and AABR showed that the implementation of UNHS for congenital permanent childhood hearing loss among all newborns in Ireland is accurate, feasible and effective. Identification of all newborns with hearing loss has now become an attainable realistic goal in Ireland. There is no reason why any child born in Ireland should experience anything other than normal acquisition of communicating skills as a result of early intervention. The implementation of the UNHS as a national program in Ireland is long overdue.

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Comments:
