Developing a Universal Newborn Hearing Screening Program: The Australian Experience and Outcomes

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Introduction

Throughout the world, there has been progressive introduction and implementation of Universal Newborn Hearing Screening programs (UNHSPs) since the first programs in the USA in Rhode Island in 1988. The incidence of significant congenital sensorineural hearing loss (SNHL) averages between 1-1.5 newborns per thousand births and there is much evidence that early detection and habilitation can have significantly improved language outcomes in those babies detected early by UNHSPs.

The development of UNHSPs has been either through a '*top down'* process as in the UK, where a national consensus group advocated and helped implement the UK UNHSP in 2000, or a '*bottom up'* process as in the USA and indeed in Australia, where a few passionate individuals from various medical, educational and paramedical fields convinced local state governments and eventually federal governments of the need for UNHSPs.

This chapter outlines the history of the development of UNHSPs in Australia, the challenges experienced and still to be resolved, and the outcomes so far in early detection, habilitation and in economic terms.

Implementation history

During the 1990s in a few birthing hospitals throughout Australia, babies in neonatal intensive care units (NICUs) with known risk factors for SNHL (**Table 1**) were screened for SNHL as 'at risk' babies utilising either oto-acoustic emission screeners or automated Auditory Brainstem Response audiometry (AABR) (**Figure 1**). However it was apparent to many Australian participants at the Newborn Hearing Screening meetings (Milan, 1998) that we were missing at least half of the babies born with SNHL without risk factors. In Western Australia, a pilot study funded by Lions Hearing Foundation and administered by an audiologist

Figure 1. Auditory Brainstem Response audiometry (AABR) testing.

and paediatric otolaryngologist, assessed approximately 1700 neonates between 1998 and 1999. This study revealed four neonates with severe to profound bilateral SNHL, two with risk factors and two without.

Table 1: JCIH risk indicators associated with permament congenital, delayed-onset, or progressive hearing loss in childhood.

Level of concern for delayed-onset hearing loss	Risk factor associated with permantent congenital delayed-onset, or progressive hearing loss in childhood
greater concern	Caregiver concern§ regarding hearing, speech, language, or developmental delay.
greater concern	Family history§ of permanent childhood hearing loss.
greater concern	Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO,§ assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion
greater concern	In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.
	Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
	Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
greater concern	Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
greater concern	Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
greater concern	Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
greater concern	Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.
greater concern	Chemotherapy.§

Risk indicators that are marked with a "§" are of greater concern for delayed-onset hearing loss.

With this evidence the Minister of Health in Western Australia, Dr John Day, funded a large scale trial of newborn hearing screening in 45% of babies born in Western Australia. 12,708 babies – 96.2% of 13,214 eligible babies – were screened between February 2000 and June 2001 utilising Transient Evoked Otoacoustic Emissions (TEOAE) or AABR. 99% of screened babies had a pass in their

initial or follow-up screening, and of the 23 babies referred for diagnostic ABR, 9 were diagnosed with bilateral SNHL (Bailey *et al*., 2002). Significant funding from the Garnett Passe and Rodney Williams Memorial Foundation helped in the administering and research outcomes of this trial, paving the way to implementation of UNHS in Australia.

In March 2001, 110 interested participants, including audiologists, teachers of the deaf, neonatologists, paediatricians, paediatric otolaryngologists, epidemiologists, nurses and parents of children with permanent hearing impairment met in Adelaide for the'Universal Hearing Screening in Australia; a National Forum for Consensus and Implementation' and promulgated a consensus statement on UNHS, a summary of which is in **Table 2**.

In 2002, New South Wales (NSW) introduced a UNHSP, closely followed by Queensland and South Australia. By 2009, the Commonwealth Government had mandated that every Australian baby should be given the opportunity to have a newborn hearing screening by the end of 2010. This lead to the other States and Territories coming on board with the target rate for hearing screening of 97% of neonates born in Australia. The target level at which the screening is performed is for permanent childhood hearing loss of 40 dB or greater and this can be bilateral, unilateral, sensorineural or permanent conductive hearing loss.

Table 2. Mission and Aims of the Australasian UNHS Committee. MISSION AND AIM

Mission To promote early detection and intervention for all Australian children with permanent hearing impairment.
1. To promote accessible and appropriate assessment, both audiological and medical, for all children who are identified by those screening programs.
2. To promote immediate and appropriate intervention for all children identified with permanent hearing impairment in Australia.
3. To promote immediate and appropriate support for all parents of children identified with permanent hearing impairment in Australia.
4. To promote the establishment of a national database of permanent childhood hearing impairment, covering severity, aetiology, age of onset and manner of detection, in every state, territory or health region of Australia, for the monitoring of outcomes and epidemiology to inform research and service provision.
5. To develop appropriate models, standards and protocols for high quality early detection and intervention for permanent childhood hearing impairment in Australia.
6. To ensure appropriate evaluation of process and outcome for all the above.
7. To promote research into the delivery and outcomes of early detection and intervention for permanent childhood hearing impairment in Australia.
8. To facilitate discussion and sharing of experience among those concerned with early detection and intervention for permanent childhood hearing impairment in Australia.

Screening occurs at birthing hospitals within the first few days of birth or, in the case of short stay babies, at a follow-up clinic. Although some Australian States used an initial TEOAE screen, at this time AABR is utilised nationwide in order to increase accuracy, and to avoid missing those babies with auditory neuropathy/auditory dyssynchrony. A neonate either passes or refers from the first screening, and the AABR screen is repeated within two weeks. If the refer result

is repeated, the neonate is referred for diagnostic testing within two months. This should mean the infant has definitive hearing testing by three months of age (actual or corrected premature). Simultaneously the infant is referred for ENT, Ophthalmic, Genetic, and General Paediatric consultation, and also referred to Australian Hearing, an non-government organisation (NGO) which supplies, amongst other services, hearing amplification to all Australian children till age 26 years, free of charge. This organisation is therefore seeing virtually all hearing impaired children in Australia and is in the unique position of being able to monitor children with hearing loss during their childhood.

In addition, children with high at risk factors such as severe jaundice at birth, CMV infection, and who are syndromic, or those with a family history of progressive SNHL have re-evaluation of their hearing at 9-12 months and subsequently if indicated.

Australian otologists also, with our pioneering multi-channel cochlear implant technology have implanted many children under the age of twelve months in order to achieve the best language outcomes for severe to profoundly deaf babies.

Key performance indicators for success in the Australian UNHSPs are as outlined in **Table 3**. Specifically the Joint Committee on Infant Hearing benchmarks, adapted by most of our State programs are:

1. Hearing screening completed in 95% of newborns by 1 month of age;
2. Referral rate to audiological assessment of less than 4%;
3. Diagnostic audiological testing is completed in 90% of cases by 3 months.

Table 3. National performance indicators for neonatal hearing screening in Australia

Performance Indicators	Aim
Participation - Participation in screening	To maximise the number of eligible infants screened for permanent childhood hearing impairment
Screening - Positivity rate of the screening test - Positivite predictive value of the screening test	To maximise the indetification of infants with potential hearing impairment while minimising parental anxiety and cost
Audiological assessment and diagnosis - Audiological assessment - Detection of permament childhood hearing impairment	To accurately identify infatns born with permanent childhood hearing impairment
Early invervention and management - Attend early internvetion service - Infants fitted with an assistive hearing devide	To maximise engagement of infants identified as requiring a service with early intervention services

Outcomes

The average age for detection of SNHL in 1997 was 25 months, while in 2015 the average age at diagnosis was 3 months. The rates of hearing aid fitting to infants under 6 months increased from 27% to 73% in the period from 2003-2009.

Approximately 3 million neonates have been tested since 2000 and some 3700 infants have been diagnosed with bilateral moderate, severe or profound SNHL. The cost savings to the Australian Government with early detection and habilitation of infants with SNHL in terms of medical, educational and vocational savings for the lifetime of these children has been estimated at almost 4 billion Australian dollars (3.06 billion US dollars), even taking into consideration the costs of hearing aids, cochlear implantation and ongoing habilitation.

Teresa Y.C. Ching PhD, a senior research scientist and head of the Rehabilitation Procedures research at the National Acoustic Laboratory in Australia, instituted the Longitudinal Outcomes of Children with Hearing Impairment (LOCHI) study to compare over 5 years the outcomes of children with hearing loss who received early or late intervention (Ching, 2013). When the 450 LOCHI children were reviewed and had their language and pre-reading skills assessed at age 5 years, those children who had been fitted with hearing aids before 6 months had better language scores than those whose hearing loss was picked up later. Similarly, those children with severe to profound SNHL who received a cochlear implant by 12 months had significantly better spoken language scores compared with those implanted at an older age. It was also noted that the hearing impaired children had significantly impaired pre-reading skills compared with their normal hearing peers. The LOCHI children are currently undergoing their 9 year assessment and this may well confirm the benefits to the child and society of the benefits of UNHS on language and academic achievements in children with hearing impairment.

Conclusion

In Australia, the 16 year experience with UNHS programs has had a dramatic effect in detecting SNHL in the perinatal period, supplying hearing aids by 6 months and Cochlear implants, if indicated, by 12 months of age. The language, academic and vocational outcomes of the UNHSPs are significant, and the cost-effectiveness and societal benefits potentially great.

Recomended readings

1. Medical Services Advisory Committee (MSAC) 2007, Universal neonatal hearing screening assessment report. Reference 17. Commonwealth of Australia.
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3. Webpage: www.outcomes.nal.gov.au
4. Schroeder L. *et al*, 2006. The economic costs of congenital bilateral childhood hearing impairment. Paediatrics 117(4): 1101-1112.
5. Dahl H-HM, Ching TYC, Hutchison W, Hou S, Seeto M, Sjahalam-King J (2013) Etiology and Audiological Outcomes at 3 years for 364 Children in Australia. PLoS ONE 8(3): e59624.doi:10.1372/journal.pone.0059624
6. Bailey HD, Bower C, Krishnaswamy J, Coates HL. Med J Aust 2002 Aug 19;177(4):180-5. Newborn hearing screening in Western Australia.
7. Australian Institute of Health and Welfare 2013. Cancer and Screening Unit Working Paper. National performance indicators to support neonatal hearing screening in Australia. Cat no.CAT 73. Canberra: AIHW.