A STUDY OF OUTCOME OF NEWBORN HEARING SCREENING PROGRAMME IN HUSM FROM JANUARY 2003 TO DECEMBER 2007

By:

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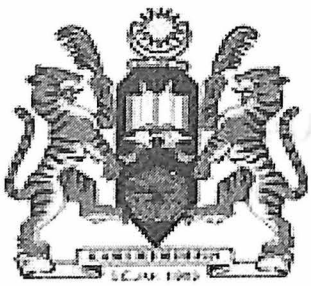

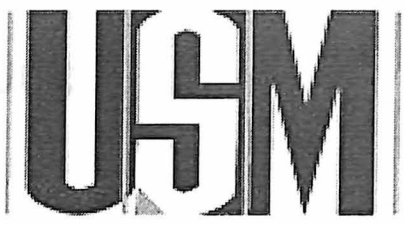

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ABBREVIATIONS

ABR	=	auditory brainstem response
AABR	=	automated auditory brainstem response
ASHA	=	American Speech-Language-Hearing Association
BAEP	=	brainstem auditory evoked potential
BAER	=	brainstem auditory evoked response
BERA	=	brainstem evoked response audiometry
BSER	=	brainstem evoked response
dB HL	=	decibel hearing level
dB SPL	=	decibel sound pressure level
DPOAE	=	distortion product otoacoustic emission
ECMO	=	extracorporeal membrane oxygenation
EHDI	=	early hearing dectection and intervention
HUKM	=	Hospital Universiti Kebangsaan Malaysia
HUSM	=	Hospital Universiti Sains Malaysia
JCIH	=	Joint Committee on Infant Hearing
NICU	=	neonatal intensive care unit
OAE	=	otoacoustic emission
SFOAE	=	stimulus frequency otoacoustic emission
SPOAE	=	spontaneous otoacoustic emission
TEOAE	=	transient evoked otoacoustic emission
UNHS	=	universal newborn hearing screening

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ABSTRACT

Universal newborn hearing screening has been started in HUSM since January 2003. The effectiveness and the challenges of the programme need to be evaluated. This retrospective study aims to determine the outcome of the newborn hearing screening in HUSM from January 2003 to December 2007. All infants who were delivered in HUSM were screened for hearing impairment with portable distortion product otoacoustic emission (DPOAE) before discharge. When they failed the initial screening, a second screening with DPOAE was arranged in 6 weeks. When these newborns failed the second screening, an audiologist performed a diagnostic auditory brainstem response (ABR) test to confirm the hearing loss. In this study, the data of 16,100 newborns were traced from the newborn hearing screening record. The data such as sex, race, age, results of the first, second and third screening were analyzed with SPSS 16.0. The study showed that the coverage of the UNHS was 98%. The prevalence of hearing impairment was 0.09%. The prevalence of initial screening refer rate was 25.5%. The prevalence of defaulters in second and third screening was 33.9% and 40.7% respectively. The age of detection of hearing impairment was 3.31 months (S.D. 0.86). The age of hearing aid fitting was 13.57 months (S.D.4.82).

ABSTRAK

HUSM telah menjalankan saringan pendengaran bayi universal sejak bulan Januari 2003. Keberkesanan dan cabaran progam ini perlulah dinilai. Kajian retrospektif ini bertujuan untuk menentukan hasil dari saringan pendengaran bayi yang telah dijalankan di HUSM dari Januari 2003 hingga Disember 2007. Semua bayi yang dilahirkan di HUSM telah disaring untuk mengesan masalah pendengaran dengan menggunakan alat "distortion product otoacoustic emission" (DPOAE) sebelum pulang ke rumah. Bila saringan permulaan gagal, temujanji untuk saringan kedua menggunakan DPOAE diberi dalam masa 6 minggu. Bila bayi gagal ujian kedua, "audiologist" menjalankan pemeriksaan untuk mengesahkan masalah pendengaran dengan menggunakan alat diagnosa "auditory brainstem response" (ABR). Dalam kajian ini, data dari 16,100 bayi diambil dari rekod saringan pendengaran bayi. Data seperti jantina, bangsa, umur, keputusan saringan pertama, kedua dan ketiga telah dianalisa menggunakan SPSS 16.0. Kajian ini menunjukkan liputan saringan pendengaran bayi universal sebanyak 98%. Kadar masalah pendengaran adalah 0.09%. Kadar kegagalan pada saringan permulaan adalah 25.5%. Kadar tidak hadir pada saringan kedua dan ketiga adalah 33.9% dan 40.7%. Umur bila masalah pendengaran dikesan adalah 3.31 bulan (S.D. 0.86). Umur bila alat bantu dengar digunakan adalah 13.57 bulan (S.D. 4.82).

CHAPTER 1

INTRODUCTION

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Hearing is one of our most precious senses. It allows us to communicate with family and friends. It also keeps us in contact with the beautiful sounds of life surrounding us. Unfortunately, most of us don't realize how important hearing is until hearing problems begin to affect our everyday lives.

Hearing in a newborn is much more precious because normal speech and language development depend upon a child's ability to hear. Hearing loss is usually described as an invisible condition because it can go unnoticed in children. The children with hearing loss or hard of hearing may be mistaken for a developmental delay or an attention deficit disorder. Without timely identification and intervention, these children may develop delayed speech and language disability as well as impaired academic achievement and social interactions or employment difficulties.

With the above important consequences to consider, early hearing detection and intervention (EHDI) programmes have been conducted all over the world. In 2000, citing advances in screening technology, the Joint Committee on Infant Hearing (JCIH) endorsed the universal screening of all infants through an integrated and interdisciplinary system of EHDI.

1.1 Prevalence of hearing impairment in newborn

Additional information about hearing loss emerges from the thousands of newborns tested, with new prevalence figures for permanent congenital sensorineural hearing loss of 1.5 to 2.2 per 1000 live births (Sharon Fujikawa, 2000). With these statistics, hearing loss is the most frequently occurring birth defect. In comparison, hypothyroidism and phenylketonuria , which are regularly screened for at birth in the developed countries, they have prevalence rates of 25 and 7 per 100,000 live births, respectively. Both of these conditions have a prevalence rate at birth that is much lower than that for hearing loss, yet hearing loss is not regularly screened for at birth (Mehl A, Thomson, 1998). It has also been estimated that two to four per 100 infants in the newborn intensive care unit will have significant hearing loss (McMurray JS, 2000).

In Malaysia, a few studies had been done to ascertain the prevalence of hearing loss among the newborns and also among the high-risk infants. Abdullah A *et al.* (2006) found that the prevalence of hearing loss among newborns delivered HUKM was 0.42% (16/3,762). Dinsuhaimi (1991) found that the prevalence of bilateral hearing loss in infants of the special care nursery was 9.46% (18/190), 5.91% (7/152) and 1.88 % (2/108) at the first, second and third visits respectively tested using ABR. In HUSM, there were 2 studies done on the prevalence of hearing loss among high-risk neonates. The first one was done by Mohd Khairi (2001), which he found that the prevalence was 1.28%. The other study was done by Khaled Saad (2002) which showed the prevalence of 0.94%.

1.2 Universal newborn hearing screening programme

Hearing loss in newborns and infants is not readily detectable by routine clinical procedures or behavioural observation, although parents often report the suspicion of hearing loss, inattention, or unresponsiveness to sound before hearing loss is confirmed (Arehart *et al.*, 1998; Harrison and Roush, 1996). It has been demonstrated that newborn hearing screening leads to earlier identification and treatment of infants with hearing loss. Studies of the newborn hearing screening programmes in the United States have demonstrated that the mean age of identification of hearing impairment has decreased from 12-24 months before the programmes were introduced, to 3-6 months since their introduction (D.C. Thompson *et al.*, 2001). Moreover the mean age at which infants receive hearing aids has been reduced from 13-16 months before the programmes began to 5-7 months following their introduction (A. Canale *et al.*, 2006). With these significant findings of the newborn hearing screening programmes, the JCIH 1994 position statement had endorsed the goal of universal detection of infants with hearing loss and encouraged continuing research and development to improve methodologies for identification of and intervention for hearing loss.

Universal newborn hearing screening (UNHS) is becoming widely accepted as the standard of care for infants in many parts of the world. Advances in screening technology have provided programmes with efficient tools. In the United States, 36 states required, through legislative mandate, that the states establish a system for early hearing detection and intervention (EHDI) beginning with mandated UNHS. Within the past year, clinical and research reports on the effectiveness, costs, and potential risks of universal newborn

hearing screening have added to the large and continuously growing body of literature on early hearing detection and intervention. Reports from both state-wide and hospital-specific screening programmes demonstrate the feasibility and effectiveness of universal newborn hearing screening for improving early identification of infants with hearing loss (Deborah Hayes, 2002).

Here are the findings of the studies of universal newborn hearing screening that were done all over the world. There are various protocols used in the studies. The methods of screening used are also differs. In Italy, De Capua B. *et al.* (2007) found that the prevalence for all hearing loss in the neonatal period was 1.78 over 1000 live births, with bilateral hearing loss in 1.42 over 1000 live births. All the hearing loss infants were diagnosed before 3 months of age and received intervention before 6 months of age. In Mexico, Hector M. Yee-Arellano *et al.* (2006) reported a total of 3066 newborns were screened (99.9%). The prevalence of sensorineural and bilateral hearing loss was of 0.65 over 1000 newborns. Seventy-three neonates (2.37%) had a risk factor for hearing impairment. The positive predictive value for sensorineural hearing loss was 71.4% and the false positive rate was 0.065%.

Doris Nekahm *et al.* (2001) had done a study in Austria and found that since UNHS was introduced in some newborn nurseries in 1995, a significant higher number of hearing-impaired children have been detected early. For the whole sample, the increase of the early detection rate was 39.9%, with a 95% confidence interval of 33.2-46.8%. For moderate hearing loss the increase is 49.2 with a 95% confidence interval of 39.6-58.8%.

In Singapore, the universal newborn hearing screening programme was implemented in 2002. It was started in KK Women's and Children's Hospital (KKWCH) which accounted for approximately one-third of deliveries (or 15,000 per year) in Singapore. The data from Singapore General Hospital (SGH) , National University Hospital (NUH) and KK Women's and Children's Hospital (KKWCH) for the period of 1 April 2002 to 31 March 2004 showed that 99.8% (36,093) were successfully screened. A total of 220 babies (0.6%) failed the screening test using OAE and AABR and were referred for diagnostic audiological evaluation. From these, 8.2% (18) refused diagnostic audiological test and a further 8.2% (18) did not complete the necessary tests. Of the remaining 184 babies (83.6%), 146 (79.3%) were confirmed to have a hearing loss. The prevalence of hearing loss was 4 per 1000 babies with 1.7 per 1000 babies being severe or profound hearing loss. The median age of diagnosis was 2.7 months. Of the 115 infants with at least moderate hearing loss, only 55 (47.8%) were identified to be at risk for hearing impairment.

In Malaysia, audiological and intervention services for the hearing impaired children are slowly developing since the early 1990s with the development of undergraduate programmes of audiology and speech language pathology. By the year 2005, there were about 90 audiologists and 90 speech language pathologists serving about 25 million populations giving a ratio of 1 audiologist /speech pathologist to 280,000 population (S.Z. Mukari, 2006). This ratio is expected to improve with the continuing increment of the audiology and speech pathology graduates every year. A few hospitals have been implementing hospital-based newborn hearing screening since early 2000s including HUSM and HUKM. The implementation of UNHS in Malaysia has been supported by

the evidence of the negative impacts of late detection of permanent congenital hearing loss and the advantages of early intervention on language, cognitive, educational and social development (C. Yoshinaga-Itano *et al.*, 2000).

Newborn hearing screening has been started in HUSM since 1st January 2003. All babies who were delivered in HUSM were screened using its own universal newborn hearing screening protocol. Protocols are necessary to provide efficient, reliable, and valid methods for the evaluation of infants in a cost-effective manner. However, the success of this screening programme requires commitment, dedicated and well coordinated teamwork between various health professionals such as obstetricians, paediatricians, otolaryngologists, audiologists, staff nurses and other personnel. This universal newborn hearing screening programme also has a lot of challenges such as costs, shortages of manpower, inadequate support services, low public awareness and the uncertainty regarding the commitment from health care practitioners but these are not impossible to overcome (Olusanya B.O. *et al.*, 2003). These concerns point to the need for close oversight of the UNHS programme and a detailed plan that includes parent education prior to birth, screening protocols, data collection and management, delivery of information to the parents, follow-up testing when necessary, prompt intervention, and connection of the child with proper agencies and schools.

Universal newborn hearing screening programme in Malaysia is still in early phase. We still need a continuous evaluation to ensure the objectives of the programme achieved successfully and the problems are dealt with effectively. From the previous study done among patients with hearing loss attending ORL clinic in HUSM, Amran (2000) found

that the mean age of first detection was at 43 month old. This is quite late, as the age of identification is highly related to better language outcomes. The percentage of defaulters was also quite high (34.4%). The reason has to be sought and a better suggestion to overcome the problems has to be made. Abdullah A. *et al.* (2006) concluded that there were a large number of defaulters and false-positive results in his newborn hearing screening study. He suggested that the newborn hearing screening programme requires adjustments to minimize the problems. Another universal newborn hearing screening study was done using DPOAE and ABR by S.Z Mukari *et al.* (2006) which showed the coverage rate, initial refer rate and return for follow-up rate were 84.64%, 11.97% and 56.97% respectively. The study also found that the average age of diagnosis was 3.56 months.

1.3 The importance of universal newborn hearing screening

Hearing is essential for the normal development of speech. Without adequate hearing-screening programs, children with significant hearing loss often are not identified until later age. The average age of identification of congenital hearing loss in the United States in 1993, according to National Institutes of Health was about 3 years old. However, after the newborn hearing screening programme, the age has dropped to 3 to 6 months (D.C. Thompson *et al.,* 2001). The ages from birth to 5 years old are regarded as the critical period for a child's language development. Furthermore, the hearing ability during the first 6 months of life is essential for the normal acquisition of language and oral speech (A.E. Carney *et al.*, 1998). This delay in identification causes significant detrimental

effects to the child, including language delays, academic delays, psychosocial difficulties, and cognitive delays. Yoshinaga-Itano C *et al* (1998) had done a study comparing the language abilities of children who were identified either prior to 6 months of age or after 6 months of age as deaf or hearing impaired and the result showed that children whose hearing losses were identified earlier demonstrated significantly better language scores than did those children identified later.

As we know from numerous studies, the prevalence of hearing loss among high-risk infants is high between 2.5% and 10% (Salamy A. et al., 1989). However, this group of infants with hearing loss comprises only 50% of newborn population with hearing loss (Mauk G. et al., 1991). Therefore, hearing screening programs that screened only high-risk neonates missed out 50% of hearing-impaired newborns. This is certainly very significant and shows how importance the universal newborn hearing screening programmes.

1.4 Joint Committee on Infant Hearing Year 2007 Position Statement

Historically, the Joint Committee on Infant Hearing (JCIH) was established in late 1969 and composed of representatives from audiology, otolaryngology, paediatrics, and nursing with interest in children with hearing loss. The first one page Position Statement was published in Paediatrics in 1971. It concluded that data at the time were inconsistent and misleading and therefore universal screening of newborn infants could not be recommended.

However, throughout its over 30-year history, the Committee explored the complexities of hearing loss and its effect on a child's development, seeking to find newer and better methods to identify and serve the infants and their families. In 1994, JCIH recommends universal screening of hearing loss before hospital discharge and stated that all infants with hearing loss be identified before 3 months of age and receive intervention by 6 months.

The latest JCIH Position Statement was published in Pediatrics (2007). The following principles provide the foundation for effective EHDI systems and have been updated and expanded since the JCIH 2000 Position Statement:

1. All infants should have access to hearing screening using a physiologic measure before 1 month of age.
2. All infants who do not pass the initial hearing screen and the subsequent rescreening should have appropriate audiologic and medical evaluations to confirm the presence of hearing loss before 3 months of age.
3. All infants with confirmed permanent hearing loss should receive intervention services before 6 months of age. A simplified, single point of entry into an intervention system appropriate to children with hearing loss is optimal.
4. The EHDI system should be family-centered with infant and family rights and privacy guaranteed through informed choice, shared decision making, and parental consent. Families should have access to information about all intervention and treatment options and counseling regarding hearing loss.
5. The child and family should have immediate access to high-quality technology,

including hearing aids, cochlear implants, and other assistive devices when appropriate.

6. All infants and children should be monitored for hearing loss in the medical home. Continued assessment of communication development should be provided by appropriate providers to all children with or without risk indicators for hearing loss.
7. Appropriate interdisciplinary intervention programs for deaf and hard of hearing infants and their families should be provided by professionals knowledgeable about childhood hearing loss. Intervention programs should recognize and build on strengths, informed choices, traditions, and cultural beliefs of the families.
8. Information systems should be designed to interface with electronic health records and should be used to measure outcomes and report the effectiveness of EHDI services at the community, state, and federal levels.

Benchmarks and Quality Indicators

The JCIH supports the concept of regular measurements of performance and recommends routine monitoring of these measures for interprogram comparison and continuous quality improvement. Performance benchmarks represent a consensus of expert opinion in the field of newborn hearing screening and intervention. The benchmarks are the minimal requirements that should be attained by high-quality EHDI programs. Frequent measures of quality permit prompt recognition and correction of any unstable component of the EHDI process.

Quality Indicators for Screening

1. Percentage of all newborn infants who complete screening by 1 month of age; the recommended benchmark is more than 95% (age correction for preterm infants is acceptable).
2. Percentage of all newborn infants who fail initial screening and fail any subsequent rescreening before comprehensive audiological evaluation; the recommended benchmark is less than 4%.

Quality Indicators for Confirmation of Hearing Loss

1. Of infants who fail initial screening and any subsequent rescreening, the percentage who complete a comprehensive audiological evaluation by 3 months of age; the recommended benchmark is 90%.
2. For families who elect amplification, the percentage of infants with confirmed bilateral hearing loss who receive amplification devices within 1 month of confirmation of hearing loss; the recommended benchmark is 95%.

Quality Indicators for Early Intervention

1. For infants with confirmed hearing loss who qualify for Part C services, the percentage for whom parents have signed an IFSP by no later than 6 months of age; the recommended benchmark is 90%.
2. For children with acquired or late-identified hearing loss, the percentage for whom parents have signed an IFSP within 45 days of the diagnosis; the recommended benchmark is 95%.

3. The percentage of infants with confirmed hearing loss who receive the first developmental assessment with standardized assessment protocols (not criterion reference checklists) for language, speech, and nonverbal cognitive development by no later than 12 months of age; the recommended benchmark is 90%.

1.5 The overview of ear and hearing

The ear is made of outer, middle and inner parts. The ear transforms sound waves into nerve impulses which are then transmitted to the brain. The outer ear collects incoming sound waves and directs them to the tympanic membrane. The middle ear houses the ossicles that connect tympanic membrane to the oval window. The ossicles transmit the mechanical energy of sound from the tympanic membrane to the fluid-filled inner ear via the oval window. As the stapes moves in and out of the oval window in response to incoming sound waves, movement of the basilar membrane in the cochlea causes bending of the stereocilia which result in sensory stimulation of the hair cells, each of which is associated with a specific frequency region. The point of maximum basilar membrane displacement for low frequencies occurs at the apex, whereas peak displacement for high frequencies occurs at the basal end.

As the basilar membrane is displaced, the nerve fibres in the auditory nerve (VIIIth cranial nerve) are stimulated and in turn convey neural information to higher levels of the auditory system and eventually to the auditory cortex. It is important to note that the

amplitude of basilar membrane displacement is determined by both the inherent mechanical (passive) properties of the membrane and by an active mechanism associated with the outer hair cells. The outer hair cells, in effect, act as cochlear amplifiers for low level signals, a product of which is the generation of "otoacoustic emissions". Although there are many more outer hair cells than inner hair cells, the fibres of outer hair cells comprise only about 5% of the auditory nerve. The inner hair cells are associated with ascending neural fibres responsible for transmitting stimulation to higher auditory centres, while the outer hair cells respond to a descending neural feedback loop that enables the inner ear to regulate its response to incoming sounds.

From the inner ear, the cochlear portion of the auditory nerve exits the modiolus and terminates at the lower brainstem. The major nuclei of the central auditory system include the cochlear nucleus, the lateral lemniscus, the inferior colliculus and the medial geniculate. At the highest levels of the auditory system, fibres radiate from the medial geniculate to the auditory cortex. It is here in the brain's temporal lobe where hearing, in the perceptual sense, actually occurs.

1.6 High risk indicators for hearing loss in newborns and infants

Firstly, the type of hearing loss can be divided into conductive hearing loss, sensorineural hearing loss and mixed hearing loss. Conductive hearing loss occurs when there are problems in the outer and middle ear such as wax, foreign body or effusion. Sensorineural hearing loss occurs when there is damage to the inner ear (cochlea) or to

the nerve pathway from the inner ear to the brain. Mixed hearing loss occurs when there is a combination of conductive and sensorineural hearing loss.

Since 1972, the JCIH has identified specific risk indicators that often are associated with infant and childhood hearing loss. These risk indicators have been applied both in the United States and in other countries and serve two purposes. First, risk indicators help identify infants who should receive audiologic evaluation and who live in geographic locations (e.g., developing nations, remote areas) where UNHS is not yet available. The JCIH no longer recommends programs calling for screening at-risk infants because such programs will identify approximately 50% of infants with hearing loss; however, these programs may be useful where resources limit the development of UNHS. Second, because normal hearing at birth does not preclude delayed onset or acquired hearing loss, risk indicators help identify infants who should receive on-going audiologic and medical monitoring and surveillance.

Joint Committee on Infant Hearing Year 2000 Position Statement has divided the high-risk indicators into 2 categories, those present during the neonatal period and those that may develop as a result of certain medical conditions or essential medical interventions in the treatment of an ill child. These indicators are:

A. Risk Criteria: Neonates (birth-28 days)

    1. An illness or condition requiring admission of 48 hours or greater to a NICU.
    2. Stigmata or other findings associated with a syndrome known to include a sensorineural and or conductive hearing loss.