

*The Health Professions
Council of South Africa*



**PROFESSIONAL BOARD FOR SPEECH, LANGUAGE
AND HEARING PROFESSIONS**

**EARLY HEARING DETECTION AND
INTERVENTION PROGRAMMES IN SOUTH
AFRICA**

POSITION STATEMENT

YEAR 2007

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I. PREAMBLE

The Professional Board for Speech, Language and Hearing Professions of the Health Professions Council of South Africa sets out the following position regarding Early Hearing Detection and Intervention Programme (EHDI) programmes in South Africa.

The Professional Board's Year 2007 Position Statement encompasses the Position on Newborn and Infant Hearing Screening and the subsequent follow-up and intervention process. The Professional Board accepts the Joint Committee for Infant Hearing (JCIH) Year 2000 Position Statement alongside the American Academy of Pediatrics (AAP) statement on Newborn and Infant Hearing Loss as the definitive guiding documents on EHDI. These documents have therefore served to guide the formulation of an EHDI framework for South Africa but within the unique contextual characteristics of the country. Therefore a critical consideration of international benchmarks was necessary in light of research from South Africa and other developing countries towards the compilation of a contextually relevant position statement.

II. RATIONALE

The Mission of the Health Professions Council of SA and its Professional Boards is to guide the professions and protect the public. This places a responsibility on the Professional Boards to ensure that excellent standards are achieved in service delivery to patients.

EHDI programmes, as proposed in this position statement, are recommended to identify, diagnose and treat newborns and infants with disabling hearing loss as early as possible to ensure optimum, cost effective solutions that enable persons to communicate effectively, allowing them to develop to their maximum potential, and thereby to secure their full participation in, and contribution to, society and the country's economy.

This initiative is grounded on the principle that an improvement in early childhood development is central to more equal opportunities (World Bank, 2005). As stated in the 2006 World Development Report, *'Evidence supports the view that investing in early childhood has large impacts on children's health and readiness to learn and can bring important economic returns later in life—often greater than investments in formal education and training.'* (World Bank, 2005). Since differences in cognitive development start to widen from a very early age, early childhood development initiatives are central to create more equal opportunities (World Bank, 2005; Department of Social Development, 2006). This is even more pronounced in the case of children born with a disability such as childhood hearing loss, since numerous studies have demonstrated the cognitive, social-emotional, vocational and financial constraints on their development compared to those without the disability (Moeller, 2000; World Bank, 2005; Yoshinaga-Itano et al. 1998). Effective EHDI programmes, in contrast have demonstrated the ability to address the inequalities caused by the developmental constraints associated with infant hearing loss (Kennedy et al. 2005; Yoshinaga-Itano, 2004). Children in such programmes are afforded the opportunities

to develop to their maximum potential, allowing them to become participating and contributing members of their communities. Comprehensive and integrated EHDI programmes are therefore accepted and proposed as the standard of care for service-delivery to newborns and infants with hearing loss.

III. THE POSITION STATEMENT

The Professional Board for Speech, Language and Hearing Professions of the Health Professions Council of South Africa advocates early detection of and intervention for, infants with hearing loss (EHDI programmes) through integrated Provincial and District service delivery mechanisms which include all relevant government, private and non-governmental organisation (NGO) role players. This must be attained by inter-sectoral collaboration with governmental departments at all levels of care, including health, social development and education, and the private sector (Department of Social Development, 2006). The goal of EHDI is to provide children with hearing loss optimal and timely opportunities to develop linguistic, literary and communicative competence in keeping with their full potential. The adverse affects of hearing loss on language and cognitive development, as well as on psychosocial behaviour are widely reported against the established and dramatic benefits of early intervention (Kennedy et al., 2005; Moeller 2000, Yoshinaga-Itano 2004). Research evidence indicates that an infant with hearing loss who receives early intervention within the first six months of life is likely to have linguistic, speech, and cognitive development comparable to normal hearing peers in contrast to persistent delays for those who are identified late (Kennedy et al., 2005; Moeller 2000, Yoshinaga-Itano 2004). Furthermore, long-term economic benefits of universal screening programmes for hearing loss indicate reduced costs for specialised education, social welfare and improved lifetime productivity for individuals with hearing loss (Yoshinaga-Itano & Gravel, 2001). Universal newborn and infant hearing screening is therefore recommended as the preferred option for public and private health care (JCIH, 2000; Lutman & Grandori, 1999; Olusanya, Luxon & Wirz, 2005).

Universal newborn and infant hearing screening is recommended using objective physiologic measures to identify congenital and early onset bilateral hearing loss. Even though initial savings may be substantial by following a risk-based screening approach the long-term economic benefits of early identification of hearing loss will be severely compromised if a universal screening model is not applied (Yoshinaga-Itano & Gravel, 2001; Yoshinaga-Itano, 2004). Diagnostic audiological and, if necessary, medical evaluations should be in progress before 3 months of age and diagnosis confirmed by no later than 4 months of age. Those infants with confirmed hearing loss should receive intervention before 6 months of age and no later than 8 months of age from health care professionals and early interventionists with experience in infant hearing loss. In addition to these initial screens, infants demonstrating risk indicators for delayed onset or progressive hearing loss must receive ongoing monitoring by caregivers informed of the risks and the communication development milestones to observe. EHDI systems must facilitate and manage this process to ensure infants and their families will have efficient and timely access to the proposed services.

The early intervention programmes must be family-centred within a community-based model of service delivery that is culturally congruent (Fair & Louw, 1999; Louw & Avenant, 2002; Swanepoel, Hugo & Louw, 2006). Professional involvement should be within an interdisciplinary team in which families assume an equal partner role based on informed choice. The goal of informed choice is to ensure that the family decision to accept or decline the screening stems from an understanding of the consequences of each course of action (Olusanya, Luxon & Wirz, 2004a). Comprehensive and unbiased information from professional, educational and consumer organisations should therefore be provided to allow families to make informed choices. The responsibility and accountability for outcomes of EHDI programmes must be instituted at community and district levels of health care integrated with early childhood development initiatives by the departments of social development and education to provide an ongoing measurement of EHDI status and development.

IV. BACKGROUND

Hearing loss is referred to as the silent, overlooked epidemic of developing countries because of its invisible nature which prevents detection through routine clinical procedures (Swanepoel, Hugo & Louw, 2005a). It is referred to as an epidemic because of its high prevalence, being the most frequently occurring birth defect, and even though it is not a life-threatening condition, failure to intervene in time renders it a severe threat to critical quality of life indicators (Mehl & Thomson, 1998; Olusanya, Luxon & Wirz, 2004b; Swanepoel, Delpont & Swart, 2004). The adverse affects of hearing loss on language and cognitive development, as well as on psychosocial behaviour are widely reported against the established benefits of early intervention. In addition to this a society is also severely burdened by hearing loss due to the extensive economic costs associated with it. Hearing loss without adequate intervention affects an individual's ability to obtain, perform in and keep a job, and it causes people to be isolated and stigmatised during the entire course of their lives (Moeller 2000; Yoshinaga-Itano, 2004). The income of individuals with hearing loss is reported to be 40 to 45% less than the hearing population in developed countries and will be even more pronounced in developing countries like South Africa, rendering those with hearing loss the poorest of the poor (Olusanya, Ruben & Parving, 2006).

This stands in stark contrast to the body of current evidence which indicates that infants enrolled in Universal Newborn Hearing Screening (UNHS) programmes are detected earlier with hearing loss and the subsequent intervention leads to linguistic, speech and cognitive development that is comparable to normal hearing peers (Kennedy et al. 2005; Yoshinaga-Itano, 2004). These facts have led to early detection and intervention for infants with hearing loss rapidly becoming the standard of care in developed countries, with a country like the USA already screening 95% of all newborns (Morton & Nance, 2006; Kennedy & McCann, 2004). No other screening programme has demonstrated the same efficacy as UNHS programmes to reduce the age of hearing loss identification and to produce similar outcomes (Yoshinaga-Itano, 2004; Kennedy et al. 2005). Beyond the benefits to individuals, long-term economic benefits of universal screening programmes for hearing loss indicate reduced costs for specialised education, social welfare and improved lifetime

productivity, quality of life and social integration for individuals with hearing loss (Yoshinaga-Itano & Gravel, 2001; Olusanya, Ruben & Parving, 2006).

Unfortunately the momentum for implementing such widespread EHDI programmes has not carried over to the developing world where two thirds of the world's children with hearing loss reside (Olusanya, Luxon & Wirz, 2004b). Although governmental and non-governmental agencies throughout developing countries have begun to initiate programmes to prevent childhood hearing loss or to offer rehabilitation, little and slow progress toward addressing hearing loss has been reported (Olusanya, 2000, Newton et al. 2001). Poor prevalence and aetiological data for hearing loss in developing countries remains an obstacle to gain support for childhood hearing loss and to plan services (Swanepoel, Hugo & Louw, 2005b). Furthermore, data reporting the mean age of hearing loss detection and intervention is virtually non-existent due to the absence of systematic or routine screening programmes in developing countries. The initial detection of hearing loss is therefore primarily passive as a result of parental concern about observed speech and language delays, unusual behaviour or otitis media complications. The detection period can start from two years old and extend well into the adolescent years (Olusanya, 2001; Russo, 2000). These realities exacerbate the impact of hearing loss on young children in developing countries and consign them to a secluded life with limited access, if any, to education and employment opportunities (Olusanya, 2005). From an ethical and human rights perspective narrowing avoidable disparities in health care, such as those evident between children with early identified hearing loss and those without, is an important and pressing imperative (Braveman & Gruskin, 2003).

Fortunately, a renewed call from developing countries to advance the plight of children with hearing loss in these regions has recently been sounded globally (Olusanya et al. 2006). In Africa the only reports, however, have been from Nigeria and South Africa which is an indication of the lack of systematic infant hearing screening programmes in the continent (Olusanya & Okolo, 2006; Swanepoel, Hugo & Louw, 2006). This is certainly true of South Africa where apart from isolated programmes in private and public health care sectors, early identification of hearing loss is not being attained (Swanepoel, 2006; Swanepoel, Delpont & Swart, 2004). A survey of the current status of EHDI services in South Africa is an important research priority to ascertain the state of South African hearing health care services to infants with disabling hearing loss (Swanepoel, Delpont & Swart, 2004). Despite challenges in developing countries like South Africa, such as the burden of HIV/Aids on health care, investing in infants and children with hearing loss towards providing more equal opportunities with their hearing peers is an important priority. Children with hearing loss have been marginalised and benefited less from past public expenditures on essential services and therefore additional costs of including them must be accepted (UNICEF, 2005). The World Health Organization's definition of health is not just the absence of disease but the complete physical, mental, and social wellbeing of an individual and therefore health beyond survival for those infants with hearing loss can only truly be accessed through early identification and intervention (Olusanya, 2005). Available resources should therefore be distributed equitably in favour of neglected non-life threatening conditions such as infant hearing loss towards a redress of past funding negligence and a holistic and integrated improvement in population health (UNICEF, 2005; Olusanya, 2006).

The South African government recognises the importance of early intervention for children in the preventative approach proposed in the *White Paper for the Transformation of the Health System in South Africa* (Department of Health, 1997). This prevention also includes preventing secondary complications, such as developmental delays in language for infants and children with hearing loss. In addition, this paper emphasises the need for Essential National Health Research (ENHR). The *White Paper on an Integrated National Disability Strategy* (1997) furthermore calls for “*early identification of impairments and appropriate interventions*” within the primary health care system, while it also announces “*free access to assistive devices and rehabilitation services... to all children under the age of six*”. The Department of Health further specifies that free health must be provided for persons with disabilities. In accord with these goals by the Department of Health the Department of Social Development has also recently produced guidelines for early childhood development services and states that all children with disabilities have the right to inclusion, integration and mainstream facilities and all other benefits enjoyed by non-disabled peers (Department of Social Development, 2006). These rights can only truly be attained and upheld through early identification and intervention for infants with hearing loss. It is clear, therefore, that South African governmental policy guidelines favour the philosophy of screening for hearing loss in infants – it is only the implementation of such policy that is left wanting.

Equal opportunities for children with hearing loss are attainable through effective EHDI programmes and a growing body of evidence suggests long-term economic benefits to initial investments in EHDI programmes (Belli, Bustreo & Preker, 2005; Kennedy & McCann, 2004; World Bank, 2005). These facts point to a moral obligation to pursue ways of delivering services to children with hearing loss for the benefit of these individuals and South African society at large. The Board for Speech, Language and Hearing Professions of the Health Professions Council of South Africa therefore issues the year 2007 Position Statement to describe the underlying principles of effective EHDI programmes and provides guidelines and benchmarks for implementing and sustaining an accountable EHDI programme.

VI. ROLES & RESPONSIBILITIES

A. Departments and agencies

The Ministries of Health, Social Development and Education are the leading role players charged with the design and implementation of early identification, assessment and education programmes for learners with disabilities in the age group 0-9 years (Department of Education, 2001). Inter-sectoral collaborations between institutions, agencies, departments and units that may be involved in the components of an EHDI programme should be involved in an integrated way assuming responsibility for particular components. A lead agency in conjunction with tertiary training institutions should be appointed to coordinate the implementation, regulation and data management for EHDI programmes in South Africa. The Department of Health’s Maternal, Child and Women’s Health unit is recommended to steer this initiative alongside other stakeholders such as the unit

for Disabilities and Early Childhood Development from the Department of Social Development.

Performance of hearing screening programmes should be audited through a formally commissioned evaluation of designated pilot programmes. Pilot programmes should include primary as well as secondary and tertiary health care contexts and should be coordinated by the Department of Health in collaboration with tertiary institutions. Academic hospital complexes present ideal contexts for pilot programmes to establish centres of excellence that may serve as a national resource in terms of research data and protocol development for other programmes.

Provincial Directorates of Finance are recommended to accept responsibility for ensuring that an adequate dedicated allocation of funds is made to enable screening to take place, using appropriate technology. Provinces must account for the implementation of policies for free health care for persons with disabilities (Office of the Deputy President, 1997). The entire implementation of the EHDI programme should however be attained through an integrated allocation of budgets by the various ministries involved. Initially for pilot projects only and based on the results expansion of programmes should be funded. A separate allocation for assistive device technology, specifically for hearing aids must also be ensured. An important aspect noted in Chapter Two of the White Paper on an Integrated National Disability Strategy (1997) under the heading "*Implementation of the RDP to Date*" is that "*Free health care for children under six years old has not always automatically been extended to include rehabilitation and the provision of assistive devices*" (Office of the Deputy President, 1997). Steps must be taken to ensure that this is implemented as an urgent priority. In addition to the funds for assistive devices and appropriate technology, funds must be allocated for the training of personnel in the use of the equipment and the administration of screening programmes at hospitals and in the community.

B. Families and Professionals

The implementation of comprehensive services for effective EHDI programmes must rely on an interdisciplinary team approach that facilitates collaborations essential for community-based early intervention services (Moodley, Louw & Hugo 2000). Essential team members are families, audiologists, paediatricians and or primary care physicians, otolaryngologists, speech–language therapists, educators, nurses, community workers, other early intervention professionals and interpreters where needed.

Collaborative hearing services that are family-centred are based on the premise that any success a child achieves will be through family intervention, and therefore the family must be an essential and equal partner in the hearing management team (Mencher et al., 2001). Preliminary results from a South African community indicate that the majority of caregivers evidenced a willingness to participate actively in the screening process which is promising for effective collaborative teamwork in which caregivers are the primary role players (Swanepoel, Hugo & Louw, 2005c). Current caregiver knowledge and awareness

of infant hearing loss and the importance of early identification is minimal and services for supportive family education, counselling and guidance should be compiled and made available (JCIH, 2000; Swanepoel, Hugo & Louw, 2005c). Additional barriers in the South African social context such as children without parents and the excessive burden on caregivers will have to be addressed in implementing EHDI programmes for families. Adapting conventional approaches towards these contextual realities is necessary and adequate support structures must be in place.

As specified by the JCIH Year 2000 position statement audiologists are central to each component of the EHDI process from identification, evaluation and auditory habilitation for infants with hearing loss. As experts in infant hearing loss audiologists serve in the capacity of programme manager supervising the EHDI programme. In terms of the hearing screening component the audiologist develops the programme according to each context's characteristics and resources, manages the programme, assesses quality, trains support personnel, coordinates services and ensures effective transition to evaluation, habilitative and intervention services. For the follow-up component, audiologists diagnostically assess infants to confirm the presence of a hearing loss, evaluate the infant's candidacy for various amplification devices and/or assistive technology, and ensure prompt referral to early intervention services. For the early intervention component, audiologists provide timely fitting and monitoring of amplification (sensory and assistive devices) in addition to education and counselling for families in their ongoing participation in the infants' development. Audiologists furthermore may provide direct habilitation services to infants and their families and also participating in the assessment of cochlear implant candidacy (JCIH, 2000).

Paediatricians and or primary care physicians serve as the advocate for the whole child's medical welfare. Paediatricians, especially for screening programmes in NICU's and well-baby nurseries, are the key professionals overseeing the infant's health and well-being. Screening programmes must therefore be developed and managed in close collaboration and partnership with paediatricians and paediatric nurses as the primary medical practitioners responsible for infants in the NICU and well-baby nurseries. Primary care physicians and nurses at PHC clinics perform this role for infants attending for immunisations.

Otolaryngologists are another essential partner in a comprehensive EHDI programme with their speciality including the identification, evaluation, and treatment of ear diseases and syndromes related to hearing loss (JCIH, 2000). Otolaryngologists can assist in the determination of hearing loss aetiology, the presence of related syndromes and risk factors. Decisions regarding medical and/or surgical treatment in cases of hearing loss are also made by the otolaryngologist and when such medical intervention occurs the otolaryngologist also becomes involved in the long-term monitoring and follow-up of the infant. The otolaryngologist is furthermore a key member in deciding on candidacy for cochlear implantation if such specialised interventions should be made available in certain cases (JCIH, 2000).

Screening personnel can include any of the previously mentioned professionals but recommended screening personnel include trained nursing staff, community health care workers, and community volunteers. The resources in each context must guide the choice of screening personnel. Hospital-based screening in the NICU and well-baby nurseries may present an opportunity for community service audiologists or speech-language therapists to conduct the screening but nurses and/or volunteers may be more sustainable in the long-run. Screening in primary health care centres where immunisations are given present community-based primary health care nurses as the frontline health professionals in the early intervention team, since they have direct contact with at-risk infants and are based at primary health care clinics that are accessible and affordable to the majority of the South African population (Moodley, Louw & Hugo, 2000). Although nurses already have many responsibilities, the gains that can be made by developmental screening (e.g. hearing screening) are so great that sustained efforts should be made to incorporate such screening into a community nurse's day (Baez, 2003). Community health workers are also a valuable resource and could be trained to conduct hearing screening in communities.

Lay volunteers have also proved to be a valuable human resource in newborn and infant hearing screening programmes. This could be of significant value in South Africa where resources are already limited and there is a lack of health care professionals who are fluent in African languages. A community volunteer can be any person as long as he/she is motivated, has literary skills and a positive, respectful attitude towards all people (McConkey, 1995). The volunteer must receive appropriate training in the screening process and technologies and hands-on training in screening infants as well as awareness of referral patterns. The training should be provided by audiologists and periodic quality assessments must be included. The training must also empower the screening personnel to educate mothers and caregivers about the importance of returning for follow-up appointments, the effect of late-identified hearing loss, and the benefits of early identification and intervention in order to ensure efficient follow-up return rates. The quality of the training will often determine the quality of the programme (McConkey, 1995).

The family-centred early intervention programme for infants with hearing loss is primarily managed by audiologists and/or speech-language therapists or other early interventionists. These professionals must provide appropriate evaluation and treatment for language, speech and cognitive-communication development in close collaboration with caregivers and educators. Education opportunities for the future must be discussed and pursued with the family towards ensuring optimal opportunities to develop to their maximum potential, allowing them to become participating and contributing members of their communities. The early interventionist therefore supports the family in stimulation of the infant's communication development, monitors the language, speech, motor, cognitive and social-emotional development of the infant and assists the family to advocate for its unique developmental needs (JCIH, 2000).

V. PRINCIPLES

The Professional Board for Speech, Language and Hearing Professions of the Health Professions Council of South Africa endorses the development of EHDI systems in South Africa that are family-centred and community based in agreement with the JCIH Year 2000 position statement. Services must be integrated and coordinated by the relevant stakeholders including the Departments of Health, Social development and Education with input from private stakeholders and NGO's. These EHDI systems must be available to all infants in a comprehensive, coordinated and timely manner. The following six principles are provided as the foundation for effective and accountable EHDI systems in South Africa.

1. All infants are afforded access to hearing screening using a physiologic measure. Screening is conducted in three contexts: at discharge from the Neonatal Intensive Care Unit (NICU) and well-baby nurseries or through the immunisation visits at Primary Health Care (PHC) clinics. Apart from those infants in the NICU, the choice of screening platform is context driven depending on the amount of home and clinic births in the district health system. These aspects constitute Universal Newborn and Infant Hearing Screening (UNIHS) in South Africa.
2. All infants are afforded access to an effective referral system once they do not pass the initial screen and any subsequent rescreen. The referral system is efficient and prompt to appropriate audiologic and medical evaluations to confirm the presence of hearing loss by 3 months of age and no later than 4 months of age for those infants enrolled in clinic-based screening programmes.
3. All infants with confirmed permanent hearing loss receive services before 6 months of age and before 8 months of age for those infants enrolled in clinic-based screening programmes. Prompt access to assistive devices is ensured and intervention services are provided within interdisciplinary programmes that are family-centred and asset-based building on informed choice and recognition of and respect for cultural beliefs and traditions of families.
4. All infants who pass the initial screen for bilateral hearing loss but who demonstrate risk indicators for progressive, late-onset bilateral hearing loss or other auditory disorders and/or speech and language delay receive ongoing monitoring by caregivers informed of the risks and the communication developmental milestones
5. Infant and family rights are guaranteed through upholding ethical practice in terms of informed choice and consent, and appropriate protection of hearing screening evaluation and intervention results in agreement with other health care and educational information.
6. Infant and family information regarding screening and possible follow-up assessments or services must be managed by integrated information systems enable to provide data for service development. Such information systems are used to measure and report the effectiveness and efficiency of EHDI services in each District Health System as well as in private hospital complexes. Collective district, provincial and national aggregates are made available to monitor the impact of EHDI programmes on public health and education. Efforts should be made to link or integrate screening and intervention data systems in order to

determine long term outcomes of children with hearing loss. The infant and family services received from individual district public and private EHDI programmes be monitored according to the data they produce to ensure compliant and accountable functioning, to determine cost-effectiveness, and continuous quality improvement.

VII. GUIDELINES FOR EARLY HEARING DETECTION AND INTERVENTION PROGRAMMES

The following guidelines are developed from existing knowledge especially from those included in the JCIH year 2000 position statement and the AAP, and preliminary contextual research conducted in South Africa. The guidelines support the six principles of the Health Professions Council of South Africa's Year 2007 position statement and provide current information on the development and implementation of successful EHDI systems in South Africa.

In agreement with the JCIH year 2000 position statement the EHDI in South Africa position statement of the Professional Board for Speech, Language and Hearing Professions of the Health Professions Council of South Africa support the concept of applying a continual process of quality improvement at each of the EHDI components to achieve the desired outcomes. This guideline therefore provides the benchmarks and associated quality indicators for each component of the EHDI system towards monitoring its compliance and outcomes. The benchmarks for EHDI programmes are the quantifiable goals or targets that can be monitored or evaluated. Since very few UNIHS programmes have been reported in South Africa the position statement includes benchmarks from the JCIH year 2000 position statement, based on existing international data but also on some preliminary reports from South Africa and other developing countries. In instances where published data aren't available suggested benchmarks are presented. The quality indicators represent a result in terms of the specified benchmark. The quality indicators should therefore be monitored with established measures of statistical practice. Once the quality indicators for a programme are not meeting the stated benchmarks closer investigation is warranted to identify and correct the process. A discussion of each EHDI principle and the specified benchmarks and quality indicators are presented below.

A. Hearing Screening (Principle 1)

1. Targeted hearing loss and targeted population

The targeted hearing loss is a permanent bilateral hearing loss of at least 40 dB averaged over the frequencies 0.5, 1, 2, and 4 kHz. This is in agreement with the European Consensus Development Conference on Neonatal Hearing Screening's position statement (Lutman & Grandori, 1999). This targeted hearing loss is to serve as a minimal criterion for screening programmes in South Africa and as resources become available a more stringent criterion may be instituted such as adopted by the JCIH of 30 to 40 dB or more in the frequency region important for speech recognition, unilaterally and bilaterally.

The question of unilateral versus bilateral hearing loss detection becomes a compromise between the effectiveness of the treatment and the costs involved. Although research indicates that unilateral hearing loss affects developmental and emotional outcomes in children (Bess et al., 1998), limited resources inevitably place a larger emphasis on identifying bilateral hearing loss above the more expensive identification of unilateral hearing loss (Lutman, 2000). It therefore becomes a matter of selecting a target disorder within the context of available resources.

Universal screening is recommended in contrast to previous recommendations of risk-based screening for contexts with limited resources. A number of different studies have reported that the at-risk population only accounts for approximately 50% of infants with congenital hearing loss (Chu et al., 2003; Davis & Wood, 1992; Watkin, Baldwin & McEnery, 1991). Furthermore, the children identified in their first year of life through targeted Newborn Hearing Screening (NHS) have a significantly higher incidence of secondary abilities (approximately 66%) than the children identified through UNHS in well-baby nurseries (approximately 30%). This means that the children presenting only with hearing loss, who have the highest potential for success, are most likely to be missed (Yoshinaga-Itano, 2004). In addition to this UNHS demonstrates better efficacy in terms of accuracy and age of identification than risk-based screening (Grill et al. 2005). Therefore even though initial savings may be substantial by following a risk-based screening approach the long-term economic benefits to early identification of hearing loss will be severely compromised if a universal screening model is not applied (Yoshinaga-Itano & Gravel, 2001; Yoshinaga-Itano, 2004). Universal screening programmes have therefore been recommended as the preferred public health care option (Olusanya, Luxon & Wirz, 2005).

In addition to these facts, since the outset expense of making screening equipment available at hospitals and/or clinics for risk-based or universal screening will be similar and only human resources required to screen infants in universal programmes will be more, a recommendation of universal screening for bilateral hearing loss is made (Swanepoel, Louw & Hugo, 2007). Utilising a unilateral pass criterion targeting bilateral losses will reduce the time in human resources required compared to a bilateral pass criterion greatly and may prove to be a more feasible intermediate solution to identify children in most need of intervention than risk-based screening (Swanepoel, Hugo & Louw, 2006). This will keep referrals and subsequent costs low compared to a bilateral pass criterion.

A unilateral pass criterion ensures that existing resources are implemented to identify bilateral hearing loss, which impacts most significantly on a child's development. By applying a unilateral OAE pass criterion, the monetary and human resource requirements for conducting follow-up evaluations would be reduced significantly (Swanepoel, Hugo & Louw, 2006). Once programmes are functioning efficiently and sufficient capacity has been generated the protocol can be reconsidered and adapted to encompass unilateral losses also.

An important recommendation pertaining to a screening protocol targeting unilateral hearing loss, is to monitor or rescreen all the infants presenting with a unilateral refer result after the initial screen. Although an infant has passed the

screen for the targeted hearing loss a unilateral refer result is a risk indicator for development of a late-onset or progressive bilateral hearing loss (Brookhouser, Worthington, & Kelly, 1994; Murphy & Radford, 2006). This will require that although a unilateral pass meets the screening criteria the opposite ear must also be screened to establish whether the child may be at risk for a late-onset or progressive bilateral hearing loss.

2. Screening contexts

Western models of newborn hearing screening in NICU's and well-baby nurseries may not be the most appropriate screening contexts across a diverse developing country like South Africa (Swanepoel, Louw & Hugo, 2007). Although Western models of NHS has proven most effective in birthing centres before the neonate is discharged, in developing countries a significant number of births occur outside hospitals (Olusanya, Luxon & Wirz, 2004b). Reports indicate approximately one third (30%) of South African children are not born in hospitals, but the actual percentage varies greatly across regions. In the Central Karoo, for example, all births were reported to have occurred in a hospital, compared to the Tambo district where 51% of births were in a hospital, 2% were in clinics, and the other 47% were home births (Statistics South Africa, 2002). The use of 6-week immunisation visits at PHC clinics as a screening platform therefore provides a way of reaching the entire population with infant hearing screening where hospital based models will not suffice. The PHC clinics are specifically suited to the delivery of community-based services and therefore provide extensive coverage of infants in South African communities (Swanepoel, Hugo & Louw, 2006). In addition to the advantage of improved coverage, repeated immunisation visits scheduled for multi-dose vaccines provide another advantage as a ready avenue to achieve acceptably high follow-up return rates. Recent pilot studies in South Africa and Nigeria report the feasibility and potential promise of implementing infant hearing screening programmes with existing primary health care structures such as the expanded programme on immunisation (EPI) (Olusanya & Okolo, 2006; Swanepoel, Hugo & Louw, 2006; Swanepoel, Hugo & Louw, 2005c). For a discussion on aspects to consider when implementing screening at PHC clinics in South Africa consult reports by Swanepoel, Hugo & Louw (2005c, 2006). The utilisation of well-baby nurseries in hospitals and immunisation visits at PHC clinics as screening platforms must be determined by each health district according to the number of hospital and homebirths in the district. Selecting and/or combining the screening platforms must aim to optimise the district screening coverage in the most cost-effective manner.

3. Programme protocol development

The team of professionals responsible for screening in the selected contexts must conduct a comprehensive review of the prevailing infrastructure of the hospital or clinic before implementation of the screening programme. Developing hospital-based screening in the NICU and well-baby nurseries should consider technology, timing of the screening relative to discharge, availability of possible screening personnel and acoustically appropriate environments, follow-up referral criteria, information management, and quality control. Clinic-based screening programme development must consider similar aspects including technology, timing of screen with first immunisation visit at 6 weeks, coordination of follow-up screens with

subsequent immunisation visits to improve follow-up return rates (Swanepoel, Hugo & Louw, 2006), acoustically appropriate environments, availability of screening personnel, follow-up criteria, access to diagnostic evaluations, information management and quality control. Reporting and management of communication must also be defined including documentation of screen outcomes on medical records (Road-to-Health card), the contents of reports to families and physicians, and methods for reporting to district health and national data sets. The methods for ensuring that communications with families are confidential, culturally sensitive, and in a language they are fluent in must also be clearly defined (JCIH, 2000).

4. Screening technologies

Only objective physiologic measures must be employed to identify newborns and infants with the targeted hearing loss. The use of a noise-emitting device such as a rattle, a whistle or any other instrument that is not an objective means of testing hearing and is therefore not endorsed for hearing screening. Such subjective screening techniques have demonstrated poor sensitivity for hearing losses other than those of a profound degree and low specificity rates, making them unreliable and inappropriate (Downs & Sterrit, 1967; Northern & Downs, 2002).

Two physiological screening technologies are endorsed including Oto-Acoustic Emissions (OAE), both distortion product (DPOAE) and transient evoked (TEOAE), and the Automated Auditory Brainstem Response (AABR). OAE's are a measure of outer hair cell functioning in the cochlea and the Auditory Brainstem Response (ABR) a measure of neural synchrony in the VIIIth nerve and lower brainstem. The sensitivity and specificity of current OAE and AABR screening methods have proved to produce low false-positive rates of 2-3%, with some reports of less than 1% (Iwasaki et al., 2004; Lutman & Grandori, 1999; Lutman, 2000; Prieve & Stevens, 2000; Spivak et al., 2000), and false-negative rates of between 6-15% as determined by studies with follow-up procedures for the entire cohort (Kennedy et al., 1998; Vohr et al., 1998; Watkin, 1996). According to Lutman (2000), both OAE and AABR techniques can achieve specificity in excess of 95%, and Colorado and Rhode Island UNHS programmes suggest screening protocols can achieve sensitivity approximating 100%.

Utilising AABR and OAE technologies to screen for the targeted hearing loss requires that interpretive criteria, based on a clear scientific rationale, for pass and refer be established for each test procedure. Automated response detection criterion are included in almost all OAE and ABR screening equipment and preferred above decision-making based on subjective interpretation. This reduces the effects of screener bias, errors on test outcome, and ensures consistency across all infants, test conditions, and screening personnel (JCIH, 2000).

Recommendations regarding screening technologies for different screening contexts are made as follows. AABR screening, although it is more expensive than OAE due to increased disposable costs, is recommended as the technology of choice for screening NICU infants before discharge (Kezirian et al. 2001; Vohr et al. 2001). The NICU population has an increased prevalence of auditory neuropathy associated with the presence of multiple risk indicators for hearing loss and since it is a neural condition it can only be identified with a neural-based

test such as the ABR (Berlin, 1999; Sinninger, 2002). In addition to this the NICU infants are often small for gestational age and/or have a low birth weight making the AABR a more suitable test for these small infants (Hall, Smith & Popelka, 2004).

OAE screening is recommended instead of AABR screening at immunisation visits (Swanepoel, Hugo & Louw, 2006). Conducting an AABR screening on infants past neonatal age becomes increasingly difficult since the babies are more restless and irritable and they become less trusting of unfamiliar personnel as they grow older (Palmu et al. 1999; Swanepoel, Hugo & Louw, 2005c). Since the AABR requires more preparation in the form of placing the electrodes and ensuring sufficient impedance, it becomes more difficult to test the infants attending the immunisation visits, whereas a simple OAE procedure requires only a probe placement, which results in a shorter average test time (Swanepoel, Hugo & Louw, 2006). Screening technologies are only however recommended at primary healthcare clinics in districts with very low hospital births as discussed in point 5 on screening protocols.

High frequency tympanometry using a 1000Hz probe tone are furthermore recommended to be available at tertiary and secondary hospitals to differentiate the aetiology of OAE refer results (Swanepoel, Louw & Hugo, 2007). The use of high frequency tympanometry has proven useful in classifying ears into different risk categories for sensorineural hearing loss and middle-ear effusion (Baldwin, 2006; Margolis et al. 2003; Swanepoel, Hugo & Louw, 2006; Swanepoel et al. 2007). Based upon these results, appropriate referrals can be made to medical personnel immediately for treating a possible middle-ear effusion or for a diagnostic audiological evaluation to save precious time during a critical developmental period in an infant's life (Yoshinaga-Itano, 2004).

5. Screening protocols

Various screening protocols for hospital-based UNHS have been implemented successfully to provide access for all newborns to hearing screening before they are discharged. Protocols vary from inpatient screening providing one or more repeat screens using the same or different technologies or outpatient rescreening within one month after discharge. The protocol must be developed within each context to maximise the follow-up return rate and minimise the number of false-positive referrals for audiological diagnosis (JCIH, 2000).

Some recommendations emerging from the current body of knowledge is made regarding protocols for the NICU and PHC clinic based screening contexts. For NICU-based screening AABR screening and rescreening before discharge is recommended as the standard of care since this is an established at-risk population with an up to 10 times higher prevalence of sensory and neural hearing losses for which the ABR is sensitive (Polinski, 2003; Suzuki & Suzumura, 2004; Yoon et al., 2003:354). In addition to this the AABR is less affected by middle-ear effusion, which NICU infants are prone to, and yields slightly better sensitivity and specificity rates for initial screens (Engel et al., 2001; Hall, Smith & Popelka, 2004).

Screening protocols using immunisation visits at PHC clinics are recommended to employ OAE technology and must be carefully planned to coincide with subsequent visits towards ensuring efficient follow-up return rates. The first immunisation visit is scheduled for 6 weeks followed by visits at 10 and 14 weeks and then 9 months. This implies that follow-up rescreens coinciding with the second immunisation visit will be within one month from the initial screen when infants are approximately 2.5 months of age. The JCIH recommends identification of hearing loss before three months of age which allows the screening schedule at 6-week visits with limited time for follow-up and confirmation of hearing loss before 3 months of age. In addition to this challenge, an infant's first immunisation may well occur any time during the first year of life even though it is scheduled for six weeks after birth and the older infants are often more difficult to test than newborns which may result in less successful infant screens (Children in 2001, 2001; Palmu et al., 1999). Fortunately the Department of Health has recently put forward a strategic plan to ensure that full immunisations are realised for all infants by one year of age (Solarsh & Goga, 2004). It remain clear, however, that a 3-month benchmark for confirmation of hearing loss in infant hearing screening programmes at PHC clinics is not attainable at present. This benchmark must therefore be extended to at least 4 months for screening programmes at PHC clinics to allow enough time across three immunisation visits (6, 10 & 14 weeks) for rescreens and diagnostic assessments. A fourth immunisation visit scheduled for 9 month may be used for monitoring infants at risk for late-onset or progressive hearing loss.

Timely and efficient confirmation of hearing loss for infants screened at 6-week immunisation visits at PHC clinics will require an integrated multi-disciplinary follow-up system. An essential component will be the education of mothers/ caregivers regarding the importance of returning for follow-up appointments, the effect of late-identified hearing loss, and the benefits of early identification and intervention. Mothers who are better educated are more likely to return for the full set of vaccinations and probably also for the follow-up hearing screenings and evaluations (Children in 2001, 2001). A large-scale initiative of this nature must, however, be carefully considered, evaluated and planned within an appropriate model of EHDI service delivery in PHC clinics (Fair & Louw, 1999).

6. Caregiver concern regarding hearing screening

The reports from developed countries are uniform in their conclusions that parental anxiety due to screening programmes is negligible and does not differ significantly from that of parents whose infant did not receive screening. In addition to this, parents of children with hearing loss demonstrate emotional availability similar to parents of children with normal hearing (Yoshinaga-Itano, 2003). In a study of parents of severely deaf children, 96% indicated that they would have wanted neonatal identification. Only a small portion indicated that they would have preferred to have waited because of the anxieties caused (Watkin et al., 1995). Clemens, Davis & Bailey (2000) in a study of 5 010 infants report that 90% of the mothers indicated UNHS to be a "good" idea, while Hergils and Hergils (2000) indicate that 95% of the parents in a study in Sweden had a positive attitude towards NHS.

According to a report by Yoshinaga-Itano (2003) neonatal identification of hearing loss through UNHS programmes does not result in greater parental stress than later-identification of hearing loss when the intervention programme contains a comprehensive counselling content. In a study of 184 parents of children with hearing loss, the parents of early-identified children were not more likely to present with stress than parents of late-identified children (Yoshinaga-Itano, 2003). Colorado data indicates that 10% of parents of infants referred for follow-up after NHS report negative emotions (Yoshinaga-Itano & Gravel, 2001). The reported stress of parents who pass the hearing screening does not prove to be significantly different from the stress reported by parents of children who have been referred for diagnostic testing. (Yoshinaga-Itano & Gravel, 2001). Preliminary data also indicates that resolution of grief by families with early-identified children occurs faster than for families with later-identified children, as long as their children develop strong language and communication skills (Yoshinaga-Itano, 2003). In general, parents report that UNHS programmes have improved their awareness of the importance of hearing, language and speech development and as a result of this exposure they can pay more attention to their child's communication skills (Yoshinaga-Itano & Gravel, 2001).

Unfortunately little data is available for caregiver perceptions of early identification of hearing loss in developing countries like South Africa. Only two published reports from Africa has provided information on maternal views on hearing loss in the region (Olusanya, Luxon & Wirz, 2006; Swanepoel, Hugo & Louw, 2005c). Results from a survey in an urban area of Nigeria agreed with reports from developed countries indicating a favourable attitude towards early detection and intervention of childhood hearing loss from mothers (Olusanya, Luxon & Wirz, 2006). A qualitative study reporting attitudes of mothers in a clinic-based screening programme demonstrated the same favourable attitude but some concerns regarding the level of awareness and knowledge of childhood hearing loss and the benefits of early detection was noted (Swanepoel, Hugo & Louw, 2005c). Developing screening programmes in South Africa, especially in rural areas should be sensitive to caregiver perceptions of childhood hearing loss and its detection which may be influenced by cultural tradition and religious beliefs (Olusanya & Okolo, 2006; Swanepoel, Hugo & Louw, 2006). Research surveys must be conducted alongside the implementation of EHDI programmes in South Africa to ascertain caregiver perceptions on hearing loss and newborn and infant hearing screening towards culturally congruent screening programmes.

7. Benchmarks and quality indicators for newborn and infant hearing screening

a) Recommended universal newborn and infant hearing screening benchmarks

Hospital-based screening:

- Within 6 months of programme initiation, hospital-based screening programmes should screen 95% of infants before discharge or before 1 month of age.
- The referral rate of the screening process for audiological and medical evaluation should be less than 5% within one year of programme initiation

- The audiologist managing the programme must document efforts to follow-up on a minimum of 95% of infants referring the initial screen. A 70% and higher follow-up return rate is considered ideal. Successful follow-up is influenced by various factors such as lack of adequate demographic information, changes in addresses or contact details, access to facilities and personal constraints such as poverty.

Clinic-based screening:

- Within 6 months of programme initiation, clinic-based screening programmes should screen 95% of infants attending their 6-week immunisation visit
- The referral rate of the screening process for audiological and medical evaluation should be less than 5% within one year of programme initiation
- The audiologist managing the programme must document efforts to follow-up on a minimum of 95% of infants referring the initial screen. A 70% and higher follow-up return rate is considered ideal. Successful follow-up is influenced by various factors such as lack of adequate demographic information, changes in addresses or contact details, access to facilities and personal constraints such as poverty..

b) Associated quality indicators of the EHDI programme screening component

Hospital-based screening:

- Percentage of newborns screened before discharge
- Percentage of infants screened before one month of age
- Percentage of infants whose screening was not done
- Percentage of infants who do not pass the hospital-based screen
- Percentage of infants with a unilateral refer result requiring a rescreen within 6 to 9 months
- Percentage of infants who do not pass the hospital-based screen who return for follow-up services (including those returning for rescreens due to bilateral referral and those with unilateral refer results returning in 6 to 9 months and those infants returning for audiological and medical evaluation)
- Percentage of infants who do not pass the hospital-based rescreen who are referred for audiological and/or medical evaluation
- Percentage of families who refuse hospital-based hearing screening
- Percentage of caregivers reporting a positive attitude toward the screening programme after the first screen

Clinic-based screening:

- Percentage of infants screened attending their 6-week immunisation visit
- Percentage of infants screened at 6 weeks of age
- Percentage of infants who do not pass the clinic-based screen
- Percentage of infants with a unilateral refer result requiring a rescreen within 6 to 9 months
- Percentage of infants who do not pass the clinic-based screen who return for follow-up services (including those returning for rescreens due to

- bilateral referral and those with unilateral refer results returning in 6 to 9 months and those infants returning for audiologic and medical evaluation)
- Percentage of infants who do not pass the clinic-based rescreen who are referred for audiologic and/or medical evaluation
 - Percentage of families who refuse clinic-based hearing screening
 - Percentage of caregivers reporting a positive attitude toward the screening programme after the first screen

The JCIH recommend that screening programmes be monitored monthly according to the quality indicators to ascertain whether a programme is meeting the expected outcomes. This allows prompt recognition and correction of any unstable component of the screening process (JCIH, 2000).

B. Confirmation of Hearing Loss in Infants Referred from UNIHS (Principle 2)

Infants in hospital-based screening programmes who meet the referral criteria for follow-up diagnostic audiologic and medical evaluations should be evaluated before 3 months of age. Infants in clinic-based screening programmes where the earliest initial screen takes place at 6-weeks of age should receive diagnostic evaluations before 4 months of age. Referrals should be for comprehensive audiologic assessment and speciality medical evaluations to confirm the existence of a hearing loss and to determine the type, degree, and if possible the aetiology of the hearing loss. Diagnostic audiologic assessments must be scheduled at the nearest facility (secondary or tertiary) with the necessary equipment to conduct appropriate assessments including diagnostic OAE and Auditory Evoked Potential equipment. Medical assessments must be referred to the nearest referring secondary health care facility. The programme manager in each district must coordinate services and address family concerns. Evaluations are described in the following sections.

1. Audiologic evaluation

Diagnosing the presence of a hearing loss and ascertaining the type and degree of the loss must be performed by a registered audiologist with experience in diagnosing infant hearing loss. An audiologic test battery including physiologic measures and developmentally appropriate behavioural techniques must be employed towards an accountable diagnosis of a hearing loss. No single test procedure may be used but a battery of tests to cross-check the results is necessary (Bachman & Hall, 1998; Jerger & Hayes, 1976). According to the JCIH (2000) '*the purpose of the audiologic test battery is to assess the integrity of the auditory system, to estimate hearing sensitivity, and to identify all intervention options*'. All audiologic assessments of young infants should provide ear-specific estimates of the type, degree, and configuration of the hearing loss.

Test-batteries for infants younger than 6 months of age should include a child and family history, an electrophysiologic measure of threshold such as ABR and/or ASSR using frequency specific stimuli, diagnostic OAEs, assessment of middle-ear functioning, acoustic reflex thresholds, observation of the infant's behavioural response to sound, and parental report of emerging communication and auditory behaviours (JCIH, 2000). Appropriate measures of middle-ear functioning include tympanometry with high frequency probe tones of 660 or 1000 Hz, but preferably

1000 Hz (Baldwin, 2006; Margolis et al. 2003; Kei et al. 2003; Swanepoel et al. 2006), bone conduction ABR or ASSR (Cone-Wesson & Ramirez, 1997; Small & Stapells, 2006) and/or pneumatic otoscopy.

The test-battery for infants and toddlers between 6 through 36 months of age should include a child and family history, behavioural response audiometry according to the child’s developmental age (visual reinforcement or conditioned play audiometry), speech detection and recognition measures, parental report of auditory and visual behaviours, and a screening of communication and language milestones (JCIH, 2000). Physiologic measures such as OAE, ABR and ASSR should also be conducted at least on the initial evaluation but also on subsequent evaluations as necessary.

The diagnosing audiologists must make a decision in consultation with the family regarding appropriate intervention options and necessary referrals based on the evaluation outcome. The audiologist managing the screening programme must schedule follow-up appointments to provide personal amplification systems and identify appropriate professionals to assess the child’s level of functioning and provide comprehensive unbiased family-centred intervention services.

2. Medical evaluation

Infants with confirmed hearing loss and/or middle ear dysfunction should be referred for otologic and other medical evaluations as needed. The purpose of these evaluations include determining the aetiology of hearing loss, identifying related physical conditions, recommending medical treatment and referral for other services as necessary. Necessary components of the medical evaluation include clinical history, family history, physical examination as well as indicated laboratory and radiologic studies. An otolaryngologist may also consult with a geneticist for chromosome analysis and evaluation of specific syndromes associated with hearing loss (JCIH, 2000).

A summary of medical professionals involved in managing the infant with a hearing loss and their responsibilities in the team is listed in table 1.

TABLE 1. Medical management of infants and children with hearing loss (Summarised from the JCIH Year 2000 Position Statement)

PHYSICIAN	RESPONSIBILITY
a) Paediatrician or primary care physician	<ul style="list-style-type: none"> - Monitoring general health and well-being of the infant - Assures the audiologic assessment is conducted on infants who do not pass their screening in partnership with family and other health care professionals - Initiates referrals for medical speciality evaluations necessary to determine hearing loss aetiology - Monitor middle-ear status because middle-ear effusion can further compromise hearing - Review risk indicators of infant and ensure periodic audiologic assessment for infants with risk factors for progressive and/or late onset hearing loss. - Monitor developmental milestones since 30- 40% of children with confirmed hearing loss demonstrate developmental delays or other disabilities (Karchmer & Allen, 1999) - Initiate referrals related to suspected disabilities

- b) Otolaryngologist**
- Evaluation including clinical history, family history, physical assessment and laboratory tests involving the ears, head, face, neck and such other systems as skin (pigmentation), eye, heart, kidney, and thyroid that could be affected by childhood hearing loss (Tomaski & Grundfast, 1999)
 - Physical examination of the ear involving identification of external ear malformations (e.g. preauricular tags and sinuses, abnormalities or obstruction of ear canals such as the presence of excessive cerumen, and abnormalities of the tympanic membrane and/or middle ear, including otitis media with effusion.
 - Supplementary evaluations may include imaging studies of temporal bones and electrocardiograms. Laboratory assessments useful in identifying aetiology may include urinalysis, blood tests for congenital or early-onset infection (e.g. cytomegalovirus, syphilis, toxoplasmosis), and specimen analyses for genetic conditions associated with hearing loss.
- c) Other medical specialists**
- A medical geneticist may be required to investigate aetiology in certain cases and to counsel families (More than 300 forms of syndromic hearing loss has been identified and for non-syndromic hearing loss, which comprises the majority of hearing loss cases, 110 chromosomal loci and at least 65 genes have been identified (Morton & Nance, 2006)
 - Other medical speciality areas may include developmental paediatrics, neurology, ophthalmology, cardiology and nephrology to determine the presence of related body-system disorders as part of syndromes associated with hearing loss.
 - Every child with hearing loss should receive an ophthalmologic evaluation at regular interval to rule out concomitant late-onset vision disorders.
 - Many infants with hearing loss will have graduated from the NICU and because these infants often demonstrate other developmental disorders the assistance of a developmental paediatrician may be valuable in management of these infants.
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3. Benchmarks and quality indicators for confirmation of hearing loss

b) Recommended benchmarks for confirmation of hearing loss

Hospital-based screening:

- Services for infants and families referred following screening are coordinated and monitored by the screening programme manager in consultation with the family
- Infants referred in the hospital-based screening programme begin audiologic and medical evaluations before 3 months of age or 3 months after discharge for NICU infants
- Infants with evidence of a hearing loss receive an otologic evaluation
- Families and professionals perceive the audiologic and medical evaluation process as positive and supportive
- Families receive support coordinated by the screening programme manager in terms of referral to appropriate intervention programmes, and provision of information to families regarding hearing loss and intervention options.

Clinic-based screening:

- Services for infants and families referred following screening are coordinated and monitored by the screening programme manager in consultation with the family

- Infants referred in the clinic-based screening programme begin audiologic and medical evaluations before 4 months of age
- Infants with evidence of a hearing loss receive an otologic evaluation
- Families and professionals perceive the audiologic and medical evaluation process as positive and supportive
- Families receive support coordinated by the screening programme manager in terms of referral to appropriate intervention programmes, and provision of information to families regarding hearing loss and intervention options.

c) Associated quality indicators of the confirmation of hearing loss

Hospital-based screening:

- Percentage of infants and families whose diagnostic evaluations is coordinated by the programme manager
- Percentage of infants whose audiologic and medical evaluations are obtained before an infant is 3 months of age
- Percentage of infants with confirmed hearing loss referred for otologic evaluation
- Percentage of families who accept audiologic and medical services
- Percentage of families of infants with confirmed hearing loss who have enrolled in an intervention programme by the time the infant is 6 months of age

Clinic-based screening:

- Percentage of infants and families whose diagnostic evaluations is coordinated by the programme manager
- Percentage of infants whose audiologic and medical evaluations are obtained before an infant is 4 months of age
- Percentage of infants with confirmed hearing loss referred for otologic evaluation
- Percentage of families who accept audiologic and medical services
- Percentage of families of infants with confirmed hearing loss who have enrolled in an intervention programme by the time the infant is 8 months of age

C. Early Intervention (Principle 3)

Early intervention for infants and young children with hearing loss has demonstrated the potential to positively influence and change cognitive and developmental outcomes in a very significant manner (Kennedy et al. 2005; Moeller 2000; Yoshinaga-Itano, 2004; Yoshinaga-Itano et al. 1998;). These benefits are attributed to the principle of critical developmental periods allowing for optimal cognitive, language and speech development that are accessed most effectively early in life. The components of the early intervention system are therefore designed to capitalise on this principle towards the development of each child's full potential.

1. Early intervention programme development

The primary member in an early intervention team is the family and therefore the programme must be designed to be responsive to the needs of each infant and his/her family. This includes addressing aspects such as the acquisition of communicative competence, social skills, emotional well-being, and positive self-esteem within a culturally congruent programme (JCIH, 2000; Louw & Avenant, 2002). Essential principles of effective early intervention are briefly described by the JCIH year 2000 position statement. In a country like South Africa, however, the characteristic linguistic, racial and cultural diversity requires the development of culturally congruent early intervention programmes (Louw & Avenant, 2002). Children acquire language within the family context where there is a dynamic interaction between language, culture, values and child rearing practices. It is therefore imperative that models of early intervention be sensitive and incorporative of the cultural-linguistic context for intervention since acquiring language and becoming a cultural member are deeply embedded processes (Crago & Eriks-Brophy, 1993; Louw & Avenant, 2002). A detailed consideration of using culture as the context for intervention for children with hearing loss is provided by Louw & Avenant (2002).

The primary health care approach adopted by the South African government requires that first world models of early intervention service delivery be adapted to adhere to the public health care philosophy of community-based primary health care (Fair & Louw, 1999). As a result, the integration of conventional early intervention models and a community-based model of service delivery as proposed by Fair and Louw (1999) should guide EHDl service delivery. The individual strengths of the two models are anticipated to be a powerful means of preventing primary, secondary and tertiary communication disorders through community participation (Fair & Louw, 1999).

In a developing country like South Africa with limited early intervention support services a 'scaling up' approach to service delivery is recommended (Olusanya et al. 2007). Successful public health programmes often start small followed by a systematic scaling up of services. Despite the fact that adequate support services in all communities are not yet available the early detection of hearing loss will provide the incentives for the systematic scaling up of services to meet the emerging and growing needs (Olusanya et al. 2007). Legislative support for early intervention services by the managerial and consultative participants in the community-based intervention process is necessary even if pilot projects are identified as an intermediate step (Fair & Louw, 1999). The responsibility for ensuring adequate allocation of funds for such projects must be negotiated between Provincial Directorates of Finance and research councils such as the Medical Research Council (MRC) and National Research Foundation (NRF) and even international organisation such as the World Bank, UNICEF and WHO. Intervention services may require setting up public-private partnerships in the initial stages towards developing more comprehensive intervention programmes for infants and children with hearing loss relying on the private and public health care systems. Ultimately however the services should be sustainable and therefore the services must be established and integrated in consultation with the government sectors.

2. Audiologic habilitation

Personal amplification or sensory device of some form is recommended for all infants and children identified with the targeted hearing loss. Families are responsible for choosing personal amplification for their infant after they have been informed regarding the various options. Audiologists are responsible to conduct the hearing aid selection and fitting in a timely fashion to minimise the amount of time between diagnosis and amplification (JCIH, 2000).

The provision of the personal amplification device should be based on physiologic threshold information if behavioural estimations are unreliable or unobtainable. Corroborating physiologic thresholds with behavioural thresholds as soon as an infant is able to provide reliable responses should be included in the follow-up schedule. The goal of amplification fitting is to provide an infant with maximum access to the acoustic spectrum of speech within a range that is safe and comfortable (JCIH, 2002). The amplification fitting protocol should include the following:

- Prescriptive procedures that incorporate individual real-ear measurements
- Validation of the benefits, particularly for speech perception, in typical listening environments
- Complementary or alternative sensory technology (FM systems, vibrotactile aids and cochlear implants) may be considered according to degree of hearing loss, goals of auditory habilitation, acoustic environments, and family's informed consent
- Long-term monitoring of personal amplification by audiologic assessment; electroacoustic, real-ear, and functional checks as well as refining prescriptive targets
- Long-term monitoring of communication, language, social emotional, cognitive and later academic development to assure that progress is commensurate with the infant's abilities.

(JCIH, 2000; Pediatric Working Group of the Conference on Amplification for Children with Auditory Deficits, 1996)

Otitis media with effusion (OME) should be promptly identified and monitored since it can further compound the sensory or permanent conductive hearing loss which reduces access to auditory/oral language stimulation and spoken language development. Referral to otolaryngologists for treatment of persistent OME is therefore indicated to ensure that amplification fitting is not delayed.

3. Medical and surgical intervention

Medical and surgical intervention required for infants with hearing loss may vary from the removal of cerumen and the treatment of OME to long-term plans for reconstructive surgery and assessment of candidacy for cochlear implants. Surgical intervention for the malformation of the outer and middle ears should be investigated in cases of permanent conductive or cases of sensory and permanent conductive losses. Further surgical intervention may be considered for infants who comply with the implantation criteria and demonstrate poor benefit from conventional amplification. The field of cochlear implants is rapidly expanding in South Africa with several implant teams around the country

(Swanepoel, 2006). Medical aids are beginning to contribute to the costs of the implants and public health care facilities have begun to implant a select few candidates who are unable to afford the device. Public-private partnerships may lead to increasing numbers of infants with hearing loss in the public health care sector receiving cochlear implants.

4. Communication assessment and intervention

Language acquisition underlies cognitive, social and emotional development in a synergistic manner (JCIH, 2000). A complete language evaluation, including oral, manual, and/or visual mechanisms and cognitive abilities should be performed for infants and young children with hearing loss. This information provides a baseline from which to support families in developing the communication abilities of their infants. Families should be provided with unbiased information specific to language development and with family-involved activities that facilitate language development in a culturally relevant manner. According to the JCIH (2000) *"the specific goals of early intervention are to facilitate developmentally appropriate language skills, enhance the family's understanding of its infant's strengths and needs, and promote the family's ability to advocate for its infant."* Families must therefore be allowed to make an informed decision regarding the communication methods including oral and visual language systems. Providing the services includes monitoring participation and progress to adapt and modify the intervention as needed. Documenting the intervention approach systematically will allow such decision making.

5. Benchmarks and quality indicators for early intervention programmes

a) Recommended benchmarks for early intervention programmes

- Infants with hearing loss are enrolled in a family-centred early intervention programme before 6 months of age for those identified through hospital-based programmes and before 8 months for those identified through clinic-based programmes
- Infants with hearing loss are enrolled in a family-centred early intervention program with professional personnel who are knowledgeable about general child development and the communication needs of infants with hearing loss
- Infants with hearing loss and no medical contraindication begin using amplification when appropriate and agreed upon by the family within one month of confirmation of the hearing loss
- Infants with amplification receive ongoing audiologic monitoring at intervals not exceeding 3 months
- Infants enrolled in early intervention achieve language development in the family's chosen communication mode that is commensurate with the infant's developmental level
- Families participate in and express satisfaction with self-advocacy
- Interaction between the multidisciplinary team serving the family (including but not limited to the audiologist, early interventionist, occupational therapist, otolaryngologist, paediatrician and physiotherapist)

b) Associated quality indicators for early intervention programmes may include:

- Percentage of infants with hearing loss who are enrolled in a family-centred early intervention program before 6 months of age for those identified in hospital-based programmes and before 8-months of age for those identified in clinic-based programmes
- Percentage of infants with hearing loss who are enrolled in an early intervention program with professional personnel who are knowledgeable about overall child development as well as the communication needs and intervention options for infants with hearing loss
- Percentage of infants in early intervention who receive language evaluations at 6-month intervals
- Percentage of infants and toddlers whose language levels, whether spoken or signed, are commensurate with those of their hearing peers
- Percentage of infants with hearing loss and no medical contraindication who begin use of amplification when agreed upon by the family within one month of confirmation of the hearing loss
- Percentage of infants with amplification who receive ongoing audiologic monitoring at intervals not exceeding 3 months
- Number of follow-up visits for amplification monitoring and adjustment within the first year following amplification fitting
- Percentage of families who refuse early intervention services
- Percentage of families who participate in and express satisfaction with self-advocacy

D. Continued Surveillance of Infants and Toddlers (Principle 4)

During the 1950s and 1960s the Hardy Group in the USA focused on the development of a list of etiological factors for sensorineural hearing loss that eventually became known as the High-Risk Register (HRR) for Hearing Loss (Mencher et al., 2000). In 1973 the JCIH recommended that mass newborn behavioural screening be discontinued in favour of testing only those infants determined to be at-risk according to five identified risk criteria on the HRR (Mahoney & Eichwald, 1987). The JCIH revised this statement in a 1982 statement when it updated the recommendations and added two more criteria to the original five high-risk indicators (JCIH, 1982). After that a number of developments led to the JCIH producing a 1994 position statement in which it changed its goal of targeted high-risk screening and endorsed *“the goal of universal detection of infants with hearing loss as early as possible. All infants with hearing loss should be identified by three months of age, and receive intervention by six months of age”* (JCIH, 1994).

Risk-based screening involves screening all newborn and infants presenting with one or more risk factor for hearing loss. This recommended screening practice subsequently evolved from risk-based to universal screening due to advances in technology and the poor yield of infants with hearing loss by high-risk screening. The discovery of the ABR in 1971 (Jewett & Williston, 1971) and OAE in 1978 (Kemp, 1978) paved the way for quasi-automatic electrophysiological NHS devices becoming available near the end of the 1980s and early 1990s (Hall, 2000; Mencher et al.,

2001). Pilot projects and continued improvements in technology demonstrated these techniques to be a fast, accurate and cost-effective means of screening newborns making UNHS a feasible possibility (Hall, 2000; Northern & Downs 2002; Roizen, 1998; Vohr et al., 1998). Furthermore risk-based screening, despite existing for decades in the USA, failed to identify a large cohort of children with hearing loss in the first year of life (Yoshinaga-Itano, 2004). Large scale studies indicated that approximately 10% of newborns and infants presented with at least one risk factor (Mahoney & Eichwald 1987; Mason et al., 1997), but this group only accounted for 45 to 50% of infants with congenital or early-onset hearing loss (Chu et al., 2003; Davis & Wood, 1992; Watkin et al., 1991). Furthermore, the children identified in their first year of life through targeted NHS have a significantly higher incidence of secondary abilities (~66%) than the children identified through UNHS in well-baby nurseries (~30%). This means that the children presenting only with hearing loss, who have the highest potential for success, are most likely to be missed (Yoshinaga-Itano, 2004). These reasons explain the replacement of targeted NHS with UNHS as the standard of care for early detection of hearing loss.

Risk indicators are still however recommended for continued surveillance of infants and toddlers and as an intermediate solution where UNHS is not immediately feasible (Olusanya, Luxon, & Wlrz, 2005). Two revised lists of risk indicators have been published in the Year 2000 JCIH position statement based on current knowledge to serve the purpose of risk-based screening and surveillance of infants at risk of late-onset or progressive hearing losses. Surveillance of infants will require that caregivers at antenatal levels of care are informed of the risk factors that these are clearly and accurately recorded on Road to Health Charts.

1. Risk-based screening (Birth through 28 days of age)

The list of risk factors specified by the JCIH year 2000 position statement is recommended for use in risk-based screening. In addition to this list, two contextual risk factors for South Africa, including maternal HIV and malaria, have been specified (Swanepoel, Hugo & Louw, 2005b). HIV has become a pandemic in South Africa with 2 in every 10 adults infected and a higher prevalence amongst females (UNICEF, 2005). The children born of HIV/AIDS infected mothers are at increased risk for hearing loss due to significantly lower birth weights, increased vulnerability for acquiring infections such as meningitis, viral encephalitis and cytomegalovirus (Spiegel & Bonwit, 2002). The direct effect of HIV exposure in-utero on newborn and infant hearing has not yet been established and requires further investigation in South Africa as a matter of priority in light of the widespread prevalence of maternal HIV. Sensorineural hearing loss may also be caused directly as a result of viral infection causing damage to the inner ear (Yoshikawa et al., 2004; Gold & Tami, 1998; Chakraborty, 2004). Viral infections may also damage the upper respiratory tract; acute otitis media and myringitis may follow with a conductive hearing loss because of the damage (Newton, 2006; Yoshikawa et al., 2004; Gold & Tami, 1998). This greater risk for developing middle-ear infections, which leads to a conductive hearing loss, may even ultimately result in a sensorineural hearing loss (Bam, Kritzinger & Louw, 2003; Matkin, Diefendorf & Erenberg 1998; Parving, 2002; Singh et al., 2003). Therefore maternal and/or infant HIV infection

presents a risk for congenital, early-onset and late-onset or progressive hearing loss. It has therefore been added to the list of risk indicators for risk-based screening and risk-based surveillance.

Malaria is furthermore added to the list of in-utero infections presenting a risk for hearing loss in South Africa. Malaria is responsible for close to three million deaths each year with one child in the world dying thereof every 30 seconds (Department of Health, 2001) and is particularly dangerous for pregnant women with the medications for treatment also being ototoxic (Claesen et al., 1998; Department of Health, 2001; Mackenzie, 2006). Many regions of South Africa are malaria prone (Department of Health, 2001) and therefore this condition was included as a risk factor unique to the South African context. Its direct effect on hearing for infants exposed prenatally has not yet been established and requires future investigation as a probable contextual risk factors for hearing loss.

The recommended list of risk indicators for South African risk-based screening is listed in table 2.

TABLE 2. Risk indicators for infants younger than 28 days

RISK-BASED SCREENING IN SOUTH AFRICA * (Birth through 28 days of age)
a) An illness or condition requiring admission of 48 hours or greater to a NICU
b) Stigmata or other findings associated with a syndrome known to include a sensorineural and or conductive hearing loss
c) Family history of permanent childhood sensorineural hearing loss
d) Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal
e) In-utero infection such as cytomegalovirus, herpes, toxoplasmosis, rubella, human immunodeficiency virus (HIV), or malaria

** Compiled from the JCIH Year 2000 Position Statement and contextual in-utero infection risks*

2. Risk-based surveillance (29 days through 3 years)

Not all infant and childhood hearing losses will be detected in the newborn. A strategy to identify acquired, late-onset, and progressive hearing losses as early as possible is an important part of a universal screening programme. These hearing losses will not be identified by newborn hearing screening and can be the result of (a) an acquired loss later in life after a traumatic event such as infection, ototoxic therapy, or chemo therapy, (b) a loss of insufficient severity to be detected by a screening procedure at birth but which progresses as the child grows, (c) a genuine late-onset loss that develops without any obvious causative factor (Fortnum, 2003). The true prevalence of such disorders is still elusive. Initial reports, based on cohorts mostly from the 1970s and 1980s in Europe, indicate that 14.5% to 27.9% of hearing-impaired children exhibit these types of hearing losses. The large range probably reflects differences in definition

(Fortnum, 2003). Reports also indicate a higher prevalence of such disorders among NICU-discharged infants (Kawashiro et al., 1996; Robertson et al., 2002).

These delayed-onset hearing losses require protocols that will ensure early identification despite having passed a newborn hearing screen. The JCIH has specified a list of risk factors for delayed-onset hearing loss to monitor infants with those risk factors for possible delayed-onset hearing loss (JCIH, 2000). Monitoring for these infants presenting with risk factors is an important priority. As UNIHS programmes continue to develop, it will become possible to determine the proportion of hearing losses in infants that are truly congenital and those that occur postnatally (JCIH, 2000). This will allow for accurate and comprehensive infant hearing screening programmes that identify congenital and delayed-onset or progressive hearing losses efficiently.

Malaria and HIV has been added to the list of risk factors for acquired, late-onset or progressive hearing losses. As stated in the previous section infant HIV and Malaria, which are common diseases in Africa may have been linked to acquired hearing loss (Sowunmi, 1997; Chukezi, 1995; Yoshikawa et al., 2004; Gold & Tami, 1998; Chakraborty, 2004). In addition to the specified JCIH list of risk indicators for acquired, late-onset, and progressive hearing losses a screening protocol targeting bilateral hearing loss must consider unilateral hearing loss as a risk factor for development of bilateral hearing loss. Increasing evidence on initial unilateral losses or unilateral refer screen results indicate a high incidence of late-onset and progressive hearing loss in the other ear leading to bilateral hearing loss (Murphy & Radford, 2006; Brookhouser, Worthington, & Kelly, 1994). Infants presenting with unilateral refer results should therefore be considered as at-risk for bilateral hearing loss.

Ideally all children at risk for bilateral hearing loss, including those with a unilateral refer result, should be scheduled for rescreens biannually for 2 years and annually thereafter until 6 years of age. Due to the reality of a health care context with constricted resources a recommendation is made that at-risk infants be monitored by their caregivers for communicative development. This will require trained personnel to inform and empower caregivers by to carefully monitor their child's hearing ability and communicative development against the milestones for normal speech and language development.

The risk factors for acquired, late-onset, and progressive hearing losses specified for risk-based surveillance in South Africa is presented in Table 3.

TABLE 2. Risk indicators for surveillance of infants and children

RISK-BASED SURVEILLANCE IN SOUTH AFRICA *
(29 days through 2 years of age)
a) Parental or caregiver concern regarding hearing, speech, language, and or developmental delay.
b) Family history of permanent childhood hearing loss.
c) Stigmata or other findings associated with a syndrome known to include a sensorineural

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- or conductive hearing loss or Eustachian tube dysfunction.
 - d) Postnatal infections associated with sensorineural hearing loss including bacterial meningitis.
 - e) In-utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis, human immunodeficiency virus (HIV), or malaria
 - f) Neonatal indicators—specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO).
 - g) Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's syndrome.
 - h) Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome.
 - i) Head trauma.
 - j) Recurrent or persistent otitis media with effusion for at least 3 months
 - k) HIV infection and Malaria

For a screening protocol targeting bilateral hearing loss:

- l) Infants with a unilateral refer result.

** Compiled from the JCIH Year 2000 Position Statement and with an additional risk indicators based on SA contextual infections and the recommended screen protocol targeting bilateral hearing loss*

E. Protection of Infants' and Families' Rights (Principle 5)

An ethical obligation resides with all professionals involved with EHDl programmes to protect the rights of the infants and families. Each institution or department involved in the EHDl process is responsible to maintain and respect these rights. These rights according to the JCIH (2000) include access to UNIHS, information in a language the family can converse in, choice, and confidentiality. The information to convey includes 1) the purpose of the screen; 2) likelihood of positive and negative findings, 3) possibility of false positive and false negative findings; 4) uncertainties and risks attached to the process; 5) any significant medical, social, or financial implications to any component of the EHDl process; 6) availability of follow-up, counselling and support services (General Medical Council, 1999; Olusanya, Luxon & Wirz, 2004b). Information should be provided in consumer-oriented language by professionals who are knowledgeable in infant hearing loss, the identification, diagnosis, and intervention process. Informed consent must be obtained before conducting any procedure as a basic legal requisite for disclosing medical information. Failure to comply is unethical and undermines the quality assurance of the EHDl process (Olusanya, Luxon & Wirz, 2004b). The family has the right to choose a preferred communication mode protected by the South African Constitution. The family has the right to confidentiality of all screening, assessment and intervention results which requires that infant and family information not be accessible in unsecured formats. Effective information management assures proper communication and confidentiality of EHDl information (JCIH, 2000).

F. Information Infrastructure and quality monitoring (Principle 6)

A national information infrastructure is vital to enable management of an EHDl programme in hospitals and community settings and to provide data for audit and

service development decisions (Waddell, 2006). Development of a national database is recommended to collate data in a uniform manner. Only such an infrastructure could facilitate effective communication between screening and intervention services. Uniform information systems are currently in use in the United Kingdom, US and Australian states. As recommended by the JCIH, this requires a standardised methodology, reporting system, and program evaluation criteria. This type of information management will serve various critical priorities including the improvement of services to infants and families; assessment of screening, evaluation, and intervention quality; compilation of data on demographics for neonatal and infant hearing loss which is currently unavailable for South Africa. A review of each of these priorities is provided in the JCIH Year 2000 position statement.

Ideally the information system must be integrated into existing systems and should maintain a record for each birth with screening, any rescreening or other assessments undertaken included. The record can include risk factor data when present as well as information on referral to early intervention. Individualized records assure that each infant receives all needed care. These recording tools should be standardised with agreement on the type of data to collect and at which level. The aggregate information from each District Health level can be integrated at each province and finally be viewed at a national level. Each District Health Department must report the number of live births and the number of newborns and infants that have been screened for hearing loss during the birth admission and first immunisation visit; the number of birthing hospitals or clinics in each district; and the number of hospitals and/or PHC clinics with universal hearing screening programmes. Other information to report is according to the quality indicators specified by the Professional Board for Speech, Language and Hearing Professions recommends that a national database should be developed.

VIII. FUTURE DIRECTIONS

In 2005, 278 million people with permanent disabling hearing loss contributed to the global burden of disease on individuals, families, communities and countries (WHO, 2005). Two thirds of these live in developing countries and 1 in every four are of early childhood onset (WHO, 2005). The significance of this health care and socioeconomic burden in childhood and its amenability to early intervention has led to revolutionary growth in newborn and infant hearing screening programmes in developed countries around the world (Morton & Nance, 2006). Infants with hearing loss in developing countries however, especially those in Africa, do not share these prospects of equal opportunities with hearing peers through EHDI programmes since an extreme dearth of early identification programmes exist (Olusanya et al., 2007). This is also true of South Africa despite a more robust health care infrastructure compared to other African countries and being the only country on the continent training audiologists. South Africa therefore has the opportunity and the moral obligation to invest in its infants with hearing loss through the implementation of widespread EHDI programmes and to take the lead in assisting other countries in Africa to provide early intervention for infant hearing loss.

To direct the future implementation of screening programmes in South Africa pilot sites must be identified for conducting consistent, evidence-based screening programmes. Pilot UNIHS programmes must be launched in NICU's, well-baby nurseries and 6-week immunisation visits in PHC clinics according to the benchmarks and quality indicators specified for these contexts in this position statement. Hospital-based pilot programmes will ideally be implemented at University health care complexes. All programmes must be jointly facilitated by the responsible Department of Health agency and research professionals at Universities with expertise in infant hearing loss. These pilot programmes must serve as centres of service excellence and as examples to other hospitals and/or clinics initiating UNIHS programmes. Furthermore, important research data concerning the efficacy of screening programmes and the contextual demographics of hearing loss will be generated at these sites towards providing contextual evidence-based data for EHDI in South Africa.

International developments in infant hearing loss that are of growing importance include employment of high frequency probe tone immittance for differential screening or diagnosis and genetic screening. Growing evidence is demonstrating higher frequency probe tones for tympanometry and acoustic reflex measurements in infants younger than 7 months is more reliable than conventional low frequency probe tones. A number of recent reports have provided guidelines and provisional norms for interpreting the more complex tympanometry shapes in these young infants (Baldwin, 2006; Kei et al. 2003; Margolis et al. 2003; Swanepoel et al. 2007). Results are promising and increasing utilisation of these tools for differentiating screen results suggesting conductive, sensorineural or mixed losses with the OAE and/or AABR results is certain.

Developments in molecular testing and identification of genetic contributions to hearing loss are an important future direction for infant hearing screening. The majority of hearing losses is attributed to genetics with an estimated contribution of 68% of congenital hearing losses and 54% for hearing losses at 4 years of age in the USA (Morton & Nance, 2006). Finding genes responsible for syndromic and non-syndromic hearing loss has been very successful with 110 chromosomal loci and at least 65 genes already identified (Morton & Nance, 2006). Moving beyond the detection of hearing loss to the identification of its cause have many potential benefits including disease prevention, improved therapy, improved interpretation of the results of early intervention and the psychological benefits of understanding the true nature of the loss (Morton & Nance, 2006). Another important and more immediate advantage of genetic screening is the identification of infants at-risk for late-onset hearing loss (Morton & Nance, 2006; NHS, 2006). Despite prevailing challenges and limitations the rapidly increasing use of diagnostic molecular testing for all infants is becoming the developed world standard of care with tests for certain genetic forms of deafness already available (e.g. GJB2 deafness and mitochondrial A1555G mutation) (Morton & Nance, 2006).

Another new development which should be considered in the future is a report that has linked a specific pattern of results on newborn hearing screening tests to sudden infant death syndrome (SIDS) (Rubens et al. 2007). This finding points towards the possibility of identifying children at risk of SIDS through the application of routine newborn hearing screening programmes. The recent report by Rubens et al. (2007)

investigated newborn hearing screening results with TEOAE retrospectively in a case-controlled study of 31 infants who subsequently died of SIDS matched with surviving controls based on gender, term versus preterm age and NICU versus well-baby nursery. Analysis of results indicated a statistically significant difference ($p < 0.05$) between the amplitude of TEOAE responses in the high frequencies (2000, 3000, and 4000 Hz) of only the right ears for infants who died of SIDS. The consistently poorer right-sided high frequency results in the experimental group was in contrast to the trend of consistently better right-sided results in the control group which is in agreement with existing literature pointing to more robust right compared to left ear results (Rubens et al. 2007). The group of SIDS infants therefore demonstrated a significant reversal of expected TEOAE results for the right ear which can easily be identified by analyzing newborn hearing screening results. The possibility of identifying infants at risk for SIDS through a simple screening technique may be an important breakthrough towards implementing preventative measures to avoid a critical incident. The association of newborn hearing screening with the detection of infants at risk for a fatal condition may serve as a breakthrough in saving lives but may also finally serve to give early intervention for childhood hearing loss in developing countries like South Africa the imperative it deserves.

IX. CONCLUSION

EHDI programmes have proved that “*hearing loss need not impede typical development, place an individual at a functional disadvantage, or alter ultimate outcome*” (Herer et al., 2002). It is time that the hearing loss barrier be minimised for children in South Africa, and that the benefits and improvement of quality of life associated with early identification and intervention become a reality for the infants who suffer hearing loss in South Africa. Children with hearing loss are as much part of the future of the country as those with normal hearing and it is through effective EHDI services that the active and equal participation of these children will be secured among their hearing peers to change, influence and direct the future of South Africa.

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