THE GLOBAL EPIDEMIC OF INFANT HEARING LOSS – PRIORITIES FOR PREVENTION

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Introduction

All children with hearing loss should be provided access to the resources necessary to reach their maximum potential (Busa et al. 2007). The first few months of life provides a critical window of opportunity for intervention to ensure optimal outcomes for infants with hearing loss. Early detection of hearing loss is the prerequisite first step in this process towards linguistic competence and literacy development (Busa et al. 2007, Korver et al. 2010, Russ, Dougherty & Jagadish 2010). Despite much success in implementing Universal Newborn Hearing Screening (UNHS) in many countries including the USA, UK, Canada and Australia more than 90% of infants with hearing loss around the globe still do not have access to the early identification services to afford them an opportunity at optimal outcomes (Olusanya 2007, Olusanya, Wirz & Luxon 2008, Swanepoel, Störbeck & Friedland 2009). This leads to poor language development, academic failure and severely restricted vocational prospects, which ultimately consign these children with hearing loss to a life of seclusion and stigmatization (Olusanya, Ruben & Parving 2006, Swanepoel 2008).

It comes as no surprise that infant hearing loss in these world regions has been coined a silent epidemic (Swanepoel 2008). It is referred to as an epidemic due to its widespread prevalence and as silent because it is not detected by routine clinical examination. Without appropriate screening caregivers may be entirely unaware that their baby may in fact have permanent hearing loss (Swanepoel 2008). It is also considered silent because it is not life threatening and as a result it does not receive the priority or visibility on global health care agendas that its long term sequela deserve. But ultimately it is a silent epidemic because the strongest advocates for
the required services - those affected by the condition themselves - are often unable to acquire sufficient spoken language and literacy to effectively promote the importance of early detection and intervention (Swanepoel 2008). This is especially true in the developing world, where more than 90% of all children under 5-years of age reside at present (UNICEF 2007).

Developed and developing – a world of inequality

The term *developing world* generally refers to countries with constrained resources and poorer performance on indicators of development such as per capita income, immunization uptake and under-5 mortality rates (World Bank 2006). Recognizing the heterogeneity of countries, the World Bank classifies them into useful groupings according to per capita income including low income, middle income (low middle and high middle income) and upper income categories. Low and middle income countries are classified as *developing* and comprise more than 5 billion people globally representing more than 80% of the global population (UNICEF 2007, World Bank 2006). Table 1 provides a comparison of selected indicators of development across country groupings globally whilst figure 1 illustrates the levels of global development across countries (IMF).

*INSERT TABLE 1 & FIGURE 1*

It is clear that there is significant variation between countries from different income groups. Mortality indicators, for example, are 10 and 20 times higher for neonatal and under-5 mortality rates, respectively, in low income compared to high income countries (WHO 2010). Life expectancy at birth varies by as much as 21 years
between low and high income regions. These health inequalities are strongly associated with inequalities in wealth with average gross national income per capita for low income countries only 3.6 % of that in high income countries. In the WHO Africa region this is most dire with 53% of the population living on less than $1 per day. These financial constraints are perpetuated in healthcare expenditure with low and lower middle income government health expenditure per capita of $11 and $34 respectively, in contrast to $2699 in high income countries (WHO 2010).

Despite these inequalities there has been an accelerated improvement in indicators such as child mortality, which is down by 30% from 1990 to 2008 with longer life expectancies at birth (WHO 2010). As improvements continue there should also be an increasing emphasis on non-communicable diseases and improved quality of life for individuals with disabilities such as hearing loss. This is especially true in the case of young children, of whom more are surviving and with longer life expectancy than ever before (WHO 2010). Early intervention can alter the entire course of their developmental, academic and vocational outcomes and ultimately reduce the burden of non-communicable diseases such as childhood hearing loss on health, education and welfare expenditure (Olusanya, Newton 2007). Nowhere are these efforts more needed than in the developing world where more than 90% of children are born (UNICEF 2007).

**Prevalence and burden of infant hearing loss**

In 1995 the WHO estimated that there are 120 million individuals with a significant (>40 dB HL) permanent bilateral hearing loss globally (WHO 2006). In 2005 this figure doubled to 278 million and if milder losses (26 to 40 dB HL) are
included, it affects almost 10% of the world population making it the most prevalent disabling condition globally (WHO 2006, WHO 2008). Childhood onset hearing loss is estimated to constitute approximately 25% of this global burden (WHO 2006). In the case of newborns, congenital or early onset permanent bilateral hearing loss affects an estimated 798,000 newborns annually (Olusanya, Wirz & Luxon 2008). At least 90% of these reside in developing countries around the world. This means that almost 2000 babies with hearing loss are born daily in developing world regions where they have no prospect of early detection and intervention services (Swanepoel, Störbeck & Friedland 2009, Olusanya, Newton 2007, Olusanya, Wirz & Luxon 2008). Apart from pilot and privately operated hearing screening programs in a few developing countries like South Africa these services are inaccessible to 9 in every 10 babies born with hearing loss (Olusanya, Wirz & Luxon 2008, Theunissen, Swanepoel 2008, Olusanya et al. 2007).

Given its prevalence and the developmental sequelae of childhood hearing loss its contribution to the global burden of disease may be significantly higher than the currently reported adult-onset hearing loss burden (Olusanya, Newton 2007, Lopez et al. 2006). Adult onset hearing loss currently ranks third on the global causes of years lived with disability (YLD) index and 15th on the disability adjusted life-years (DALY) index - only one of four non-fatal conditions among the 20 leading contributors to the global burden of disease (WHO 2008, Swanepoel et al. 2010). Without access to early intervention infants with hearing loss are assigned to a life of deprived language development, restricted academic prospects, limited literacy and poor vocational outcomes (Olusanya, Newton 2007). Considering improvements in life expectancy childhood onset hearing loss has significant implications for long-
term economic costs not only to individuals and families but on communities and countries (Olusanya, Ruben & Parving 2006, Swanepoel 2008, Olusanya, Newton 2007, WHO 2006).

Prevention of infant hearing loss in developing countries

Prevention of childhood hearing loss in developing world regions has historically been solely focused on primary prevention (Olusanya 2007, Olusanya 2005). It is only in recent years that secondary prevention services have been highlighted as an important strategy to alleviate the burden of infant hearing loss in these regions (Swanepoel, Störbeck & Friedland 2009, Olusanya et al. 2007).

Primary prevention

Addressing preventable causes of childhood hearing loss are of key importance in global efforts to reduce the burden of hearing loss particularly in developing countries where preventable cause such as infectious diseases, poor antenatal and perinatal health services and other environmental causes are widespread (Swanepoel 2008, Olusanya 2009). Table 2 lists some of the common environmental risks that may be encountered in developing regions.

*INSERT TABLE 2*

Infectious diseases like meningitis, measles and rubella are still major contributors to childhood hearing loss particularly in developing countries. Despite continual improvements in immunization coverage a disease like measles is still rife in many developing countries where almost all of the annual 25 to 30 million infected children
reside (UNICEF 2010). Widespread immunization has eliminated the risk of congenital rubella syndrome, which is closely associated with hearing loss, in many world regions but developing countries especially in sub-Saharan African still do not routinely offer rubella vaccinations (Swanepoel 2008, WHO 2010). This means that many infants may present with hearing loss secondary to congenital rubella syndrome since it is the most common sequela of this condition (Swanepoel 2008). Meningitis is probably the major contributor to acquired childhood hearing loss in developing countries especially in sub-Saharan Africa where thousands die annually but even more are left with severe disabilities including hearing loss (Swanepoel 2008, Butler 2010). A recent drive to roll-out new meningitis vaccines is offering hope that the rampant state of this disease, especially in the meningitis belt across sub-Saharan Africa, may be prevented (Butler 2010). As a result fewer cases of hearing loss may be anticipated through this primary prevention strategy (Butler 2010).

Other widespread infections such as HIV and Malaria may add to the burden of childhood hearing loss, if not directly, certainly indirectly by nature of the sheer volume of infected individuals. For example, in 2008 there was an estimated 243 million cases of malaria which caused 863 000 deaths of which the majority was children under 5 years of age (WHO 2010). Cerebral malaria is an established risk factor for hearing loss and considering the large number of cases globally many children may be expected to present with hearing loss as a result (Sowunmi 1997, Chukuezi 1995). In addition congenital malaria and its possible effect on the hearing of a developing foetus has been reported as a possible risk for hearing loss but very little is known about this to date (Olusanya, Luxon & Wirz 2004a). The HIV/AIDS pandemic poses another risk with an estimated 33 million persons infected globally -
the majority residing in sub-Saharan Africa (UNAIDS/WHO 2009). Although maternal HIV does not seem to directly cause congenital hearing loss (Olusanya, Afe & Onyia 2009) the indirect consequences of immune suppression leads to more secondary infections which may result in congenital or acquired hearing loss (Stearn, Swanepoel 2010). Furthermore, the exposure to ototoxic medications, including antiretroviral drugs, in HIV/AIDS patients pose a further possible risk for acquired childhood hearing loss (Stearn, Swanepoel 2010).

Apart from different infectious disease profiles other environmental risks in developing countries may also be very different from those encountered in developed countries. For example, undernourishment in infants is almost exclusive to developing countries and has recently been demonstrated to pose a significant risk for severe-to-profound sensorineural hearing loss early in life (Olusanya 2010b, Olusanya). Whether due to associated intra-uterine growth retardation maternal nutritional status or insults soon after birth more than half of infants with permanent hearing loss were undernourished in a study from Nigeria (Olusanya 2010b, Olusanya).

Other risk factors unique to developing world regions are the lack of skilled birth attendants with only 43% of births in low income countries assisted by skilled attendants compared to 99% in high income countries (WHO 2010). This has been highlighted as a significant risk factor for hearing loss in a recent study from Nigeria (Olusanya, Somefun 2009a). High blood pressure during pregnancy, widespread in sub-Saharan Africa, is another risk factor that is associated with a threelfold risk for sensorineural hearing loss in infancy (Olusanya, Afe & Solanke 2009). Low birth
weight, which is twice as prevalent in low and lower middle income countries compared to high income countries (Table 1), is another risk factor for hearing loss specifically relevant in developing countries (WHO 2010). A common risk factor for sensorineural hearing loss and auditory neuropathy, hyperbilirubinemia, is also more prevalent in developing countries due to a higher incidence of glucose-6-phosphate dehydrogenase deficiency (Olusanya, Somefun 2009b, Cappellini, Fiorelli 2008).

Environmental risks for infant hearing loss such as birth trauma, asphyxia and exposure to dangerous doses of ototoxic medication are also more likely to occur in lower income countries where maternal and antenatal care is lacking.

It is clear that in developing world regions the risks for childhood hearing loss are more prevalent and the risk profile may vary significantly from developed countries. The widely used list of risk factors recommended by the Joint Committee of Infant Hearing 2007 Position Statement (Busa et al. 2007) may therefore not be entirely appropriate. These risk factors are essentially hospital-based and reflect the reality of developing world health care systems (Olusanya 2010a). Neonatal intensive care unit (NICU) stay of five days or more has replaced many of the previously recommended risks such as low birth weight, asphyxia, hyperbilirubinemia requiring transfusion, mechanical ventilation lasting five days or more and ototoxic medications. The reality in many developing world regions, however, is that advanced health services such as an NICU are unavailable making this risk factor irrelevant (Olusanya 2010a). In fact the list of risk factors recommended in the 1994 Position Statement by the Joint Committee is more appropriate for developing world regions where NICU’s are not available (JCIH, 1995)
Secondary prevention

Despite a better understanding of risk profiles for childhood hearing loss in various world regions and improvements in primary prevention, up to 50% of all infant hearing losses are unrelated to these environmental risks and constitute genetic conditions (Olusanya, Newton 2007, Smith, Bale. & White 2005). Without secondary prevention efforts to identify infants early through systematic infant hearing screening programs, hearing loss will only be detected after critical language development periods have passed resulting in severely restrict prospects for literacy, academic and vocational outcomes (Olusanya, Newton 2007).

In reality, detection of infant hearing loss in developing countries remains a passive process and occurs as a result of concerns regarding observed speech and language delays or unusual behavior (Theunissen, Swanepoel 2008, Swanepoel, Störbeck & Friedland 2009, Olusanya, Luxon & Wirz 2004b). Initial detection and intervention ages for children with hearing loss in sub-Saharan Africa is well beyond three years of age due to the lack of early detection programs. The median age for hearing loss identification in a report from Angola was 6 years and ranged from between 1 to 15 years of age (Bastos et al. 1990). The mean age of first detection in a study from Kenya was 5.5 years (Omondi et al. 2007) and in Nigeria parental suspicion occurred between 12 to 24 months of age with an average 18 month delay until confirmation of hearing loss (Olusanya, Luxon & Wirz 2005). In South Africa reports from two provinces indicated the average age of first hearing loss diagnosis to be 23 and 31 months of age with the average age of hearing aid fitting at 28 and 39 months of (Swanepoel, Störbeck & Friedland 2009, Van Der Spuy, Pottas 2008).
Newborn and infant hearing screening is the only way to ensure that infants with congenital and early onset hearing loss are detected early enough to access the critical developmental period within the first year of life through intervention (Korver et al. 2010). Although the World Health Organization encourages countries to increase prevention efforts and improve access to early detection services and health surveillance systems (WHO 2010), very few developing countries have any systematic newborn or infant hearing screening programs. A recent survey of newborn and infant hearing screening in the South African public health care sector indicated that only approximately 7.5% of hospitals provide some form of infant hearing screening (Theunissen, Swanepoel 2008). If this reflects the situation in the most resourced country in sub-Saharan Africa and the only one offering a professional qualification in audiology, the situation is expected to be much worse in the majority of developing countries where there are no formal training in audiology (Goulios, Patuzzi 2008, Fagan, Jacobs 2009).

The widespread lack of any early detection programs in developing countries is despite recent studies demonstrating that early detection programs for infants in these world regions may in fact be feasible and viable (Olusanya, Wirz & Luxon 2008, Olusanya, Wirz & Luxon 2008, Olusanya et al. 2007, Swanepoel et al. 2007, Olusanya et al. 2009). Community-based screening incorporated with immunization schedules offer a way to reach infants even in countries where the majority of births may not be hospital-based (Swanepoel, Hugo & Louw 2006, Swanepoel, Louw & Hugo 2007, Olusanya, Okolo 2006). In fact, a cost analysis revealed that community-based screening programs may be significantly less expensive than hospital-based programs (Olusanya et al. 2009). In Nigeria it cost US$ 2765 to identify a child with
permanent congenital or early onset hearing loss in a hospital-based universal screening program compared to US$ 602 in the universal community-based program (Olusanya et al. 2009). Interestingly this study reported that it was more expensive to conduct targeted compared to universal infant hearing screening irrespective of hospital or community contexts (Olusanya et al. 2009). Community-based universal infant hearing screening was therefore recommended as the most cost-effective and efficient model for low-income countries. The heterogeneous nature of developing countries and even within countries will dictate what early detection approach will be most appropriate. It is clear however that early detection of hearing loss, using physiological screening techniques, should be prioritized in developing countries.

**Priorities for progress**

Charting a course towards widespread early detection in developing countries is not a simple task. It requires careful consideration alongside other health care concerns and within the limitations of available resources. Priorities and initiatives will vary greatly across the contextual, demographic and health care characteristics of countries and even within countries. But what is clear is that early detection of infant hearing loss must be prioritized for the sake of the infant, their families, and the long term economic burden associated with late identified permanent congenital and early-onset hearing loss. Priorities must include both a macro top-down approach and a micro bottom-up approach.

Global health agendas must appreciate the burden of hearing loss in infants and children. At present childhood hearing loss is not included in the calculations for the global burden of disease report (Olusanya, Newton 2007). This omission was
justified by attributing childhood hearing loss as a sequelae of congenital conditions, infectious disease or injuries (Lopez et al. 2006). In reality however, at least 50% of childhood hearing loss is genetic and many other major causes such as rubella, CMV, toxoplasmosis, mumps, herpes, neonatal jaundice and ototoxicity were all excluded from the report (Swanepoel 2008, Olusanya, Newton 2007, Lopez et al. 2006). This neglect of childhood hearing loss has meant forestalled worldwide attention and funding from global health agencies towards early detection and intervention services globally (Swanepoel 2008). Early detection and intervention for infant hearing loss should be integrated as part of maternal and child health initiatives supported by global and national health care agencies.

International collaborations should be pursued between professional associations, global health agencies and national governments to prioritize several key areas necessary for developing early detection and intervention services in developing countries. Advocacy on national and international health forums is necessary to state the case for the silent epidemic. At a local level awareness amongst health care providers regarding infant hearing loss, which is often quite poor, must be fostered (Olusanya, Roberts 2006). The lack of hearing health care personnel in developing countries requires collaborative initiatives for training and education of both specialist and non-specialist hearing personnel (Goulios, Patuzzi 2008). Utilization of the rapidly expanding network of information and communication technology in developing world regions is offering novel solutions for training and health provision through telemedicine, eHealth and mHealth models (Swanepoel et al. 2010, Swanepoel, Olusanya & Mars 2010, McCarthy, Muñoz & White 2010). Another priority for global collaborations is the possibility of leveraging economies of scale to
reduce equipment costs necessary for screening and diagnosis of hearing loss in developing countries (Olusanya 2008).

Finally, pilot programs must be initiated in developing countries in both hospital and community-based settings according to the contextual demands. These programs provide the opportunity to assess protocols and technologies whilst generating important epidemiological data for infant hearing loss in countries where such information is largely unavailable. It also inadvertently increases the awareness amongst health providers that infant hearing loss is an important health care concern and that it can be identified early with dramatic benefits if intervention is commenced as early as possible. Finally, these programs should become centers of excellence that provide families with access to necessary services, but even more importantly, to facilitate the initiation of other programs built on the successes and failures of pilot sites.

**Conclusion**

Considering that more than 90% of infants with permanent hearing loss globally are unable to benefit from early detection and intervention services creates a sense of urgency to bring services to all infant with hearing loss. Primary intervention efforts in these countries can reduce the infectious disease burden associated with congenital and early-onset hearing loss along with other environmental risks but without secondary intervention through early detection, those with hearing loss are consigned to a life of seclusion, limited access and poor quality of life (Swanepoel 2008). Addressing this silent epidemic is not simple and will require international collaborations in various areas to secure early detection and intervention services for
infants around the globe. Although daunting, the inequality of current services for infants with hearing loss raises a moral obligation to truly pursue optimal outcomes not only for a small minority of infants with hearing loss but truly for all. Those in the field of childhood hearing loss know how precious each passing month is in the life of an infant with hearing loss. Progress is being made but much more must be done much faster.

References


### Table 1. A selection of global socio-economic and health indicators (WHO 2010)

<table>
<thead>
<tr>
<th>INDICATOR</th>
<th>DEVELOPING COUNTRIES</th>
<th>DEVELOPED</th>
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<tbody>
<tr>
<td></td>
<td>Low income</td>
<td>Lower Middle income</td>
</tr>
<tr>
<td>No. countries</td>
<td>40</td>
<td>56</td>
</tr>
<tr>
<td>Life expectancy at birth 2008</td>
<td>56</td>
<td>65</td>
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<tr>
<td>Neonatal mortality rate (per 1000 live births) 2008</td>
<td>37</td>
<td>29</td>
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<tr>
<td>Low birth weight newborns (%) 2008</td>
<td>15</td>
<td>17</td>
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<tr>
<td>Under-5 mortality rate (per 1000 live births) 2008</td>
<td>109</td>
<td>36</td>
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<tr>
<td>Measles immunization coverage 2008</td>
<td>76</td>
<td>82</td>
</tr>
<tr>
<td>Median population age 2008</td>
<td>21</td>
<td>27</td>
</tr>
<tr>
<td>Aged under 15 (%) 2008</td>
<td>38</td>
<td>28</td>
</tr>
<tr>
<td>Annual growth rate</td>
<td>2.1</td>
<td>1.3</td>
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<tr>
<td>Total fertility rate (per woman) 2008</td>
<td>4.0</td>
<td>2.5</td>
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<tr>
<td>Living in urban areas (%) 2008</td>
<td>29</td>
<td>41</td>
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<tr>
<td>Per capita government expenditure on health (US$) 2007</td>
<td>11</td>
<td>34</td>
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<tr>
<td>Gross national income per capita ($) 2008</td>
<td>1372</td>
<td>4363</td>
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<tr>
<td>Population living on &lt;$1 a day (%)</td>
<td>48</td>
<td>26</td>
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### Table 2. Major infectious disease and environmental risk for hearing loss in developing countries

<table>
<thead>
<tr>
<th>Infectious diseases</th>
<th>Environmental risks</th>
</tr>
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<tbody>
<tr>
<td>Measles</td>
<td>Birth trauma</td>
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<td>Meningitis</td>
<td>High blood pressure</td>
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<tr>
<td>Rubella</td>
<td>Low birth weight</td>
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<tr>
<td>CMV</td>
<td>Neonatal jaundice</td>
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<tr>
<td>HIV</td>
<td>Undernutrition</td>
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<tr>
<td>Malaria</td>
<td>Untrained birth attendant</td>
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</tbody>
</table>
Figure 1. Classification of countries according to economic development (IMF).