Neonatal hearing screening and intervention in resource-limited settings: an overview

Bolajoko O Olusanya

ABSTRACT
From a developmental perspective, optimal speech and language outcome is indisputably the primary motivation for neonatal hearing screening of infants with congenital or early-onset hearing loss (PCHL). This paper additionally outlines more broadly the potential value of early hearing detection and intervention in resource-poor countries against the backdrop of limitations of primary prevention of PCHL based on a review of literature from low-income and middle-income countries with per capita incomes of approximately US$6000 or less. It establishes the scientific and developmental foundation for priority consideration for neonatal hearing screening and intervention in any global initiatives for effective early childhood development programmes in resource-limited countries. It also highlights approaches to addressing the various challenges to implementing effective early hearing detection and intervention programmes, and concludes with a discussion on the pivotal role of paediatricians in facilitating timely referral for requisite tests and (re)habilitative services especially for infants with established risk factors.

INTRODUCTION
Of the 122.9 million babies born annually in developing countries, about 737 000 or 6 per 1000 live births are likely to have permanent congenital or early-onset hearing loss (PCHL) compared with 28 000 or 2 per 1000 live births in developed countries. PCHL in neonates is an invisible or hidden handicap that usually becomes evident after the first year of life from delayed speech and language if systematic efforts are not made to identify and support the affected infants shortly after birth. Optimal speech and language development is crucial for psychosocial and cognitive skills and offers greater prospects for high educational and vocational attainment than non-verbal communication modes. It is therefore the most prominent effectiveness measure for early hearing detection and intervention programmes.

Empirical evidence concerning the potential benefits of early detection of PCHL for speech and language development following neonatal hearing screening programmes (NHSP) have been extensively addressed in a related paper in this issue. However, it is essential to provide additional context for evaluating the benefits of NHSP in developing countries based on its implications for the developmental trajectory of infants with PCHL in such settings. A review of evidence on the challenges and prospects for implementing effective early hearing detection and intervention is therefore necessary to inform nascent policy initiatives on early childhood development (ECD) for this region. These and related issues are examined in this paper.

DEVELOPMENTAL PERSPECTIVE OF PCHL
Global health interest in establishing the vital but often overlooked link between child survival and ECD is emerging steadily with the countdown to the 2015 target date for the Millennium Development Goals. Perhaps the most robust scientific foundation for ECD is conceptualised by the work of Thompson and Nelson on human brain development (figure 1). Broadly, human development is heralded by the formation of neural cells in the brain followed by a sequence of cell migration and differentiation in utero; from the fourth month of life myelination of peripheral nerve fibres gradually occurs to facilitate conduction from the nerve to the target organ and vice versa. Rapid brain growth follows, with neurogenesis, axonal and dendritic proliferation, synaptogenesis, cell apoptosis, synaptic pruning, myelination and gliogenesis until maximum growth is attained by the first 2 years.
Figure 1  Window for optimal auditory stimulation in human brain development. Reproduced with permission of Elsevier.  

As shown in figure 1, auditory stimulation begins in utero and peaks around the sixth postnatal month, with the fastest growth occurring within the first 3 months. Recent experimental studies have also demonstrated that early auditory stimulation is the foundation for optimal speech and language development in the first year of life. In contrast, the window of opportunity for optimal cognitive and psychosocial development extends throughout early childhood (figure 1). Ensuring that no child is at risk of poor auditory stimulation during this ‘critical’ or ‘sensitive’ period should undoubtedly be the priority of any effective ECD programme. Thus, child disability data restricted to 2–9 year olds will result in considerable underestimation of this crucial component of ECD.  

BURDEN OF PCHL IN RESOURCE-POOR COUNTRIES
In this paper, all categories of PCHL above 30 dB HL in at least one ear are considered as sufficiently significant to put an infant at risk of functional impairment based on the International Classification of Functioning, Disability and Health for Children and Youth by WHO. The lifetime impact of PCHL in resource-limited countries is usually exacerbated by adverse societal perception towards childhood disability and lack of provision within the existing health and educational systems. For example, it is not unusual for PCHL to be attributed to supernatural causes because of widely held superstitious beliefs. This fosters recourse to religious rites or unorthodox and potentially harmful traditional therapies such as medicinal plants and animal fat. Affected infants are also frequently at risk of social isolation, neglect, abuse or infanticide. Without NHSP, parents are often first to suspect a hearing impairment from a child’s inattention, erratic response to sound or speech delays. In one report from Nigeria, parents were predominantly the first to suspect or detect hearing difficulty in their children (81%), and this occurred mostly in the second year of life. Only 12% of parents suspected hearing difficulty within the first 6 months of life. The commonest mode of detection was the child’s failure to respond to sound (49%). Speech and language defects or unintelligible speech were much less likely to be associated with hearing difficulty by parents (1%). Similar findings have been reported from other developing countries. It is not unusual for the early signs of hearing impairment to be misconstrued initially as self-limiting or constitutional developmental delays only to be replaced with a deep sense of personal grief and hopelessness when a lifetime disability becomes evident. The inevitable recourse to a deaf school where sign language is the only mode of communication is in itself a source of emotional stress and anxiety for hearing parents because of the personal demand for time and resources, the prevailing sociocultural stigma and consequent isolation. As they grow into adulthood, the survivors are also likely to be restricted to a lifetime dependence on charity for basic sustenance because social benefit schemes are non-existent. The economic and social costs of undiagnosed and untreated PCHL are therefore higher in resource-poor nations than in more developed economies.

Thus the benefits of early hearing detection potentially extend well beyond achieving satisfactory speech and language outcomes. Early detection provides a rare opportunity to arrest as far as practicable the dire lifetime developmental trajectory typically associated with infants with PCHL. As parents become more knowledgeable about this invisible handicap in their apparently healthy infant they are empowered to make truly informed choices and prepare for its inevitable manifestations as the baby grows older. Such empowerment is essential for subsequent family-centred intervention services for the beneficiaries of early detection. It will help to minimise unrealistic developmental expectations of the child. Early detection also forestalls costly and time-consuming ‘diagnostic odyssey’ of trying to unravel the nature and cause of a suspected disorder and its attendant stigma. Perhaps more importantly on the supply side is the impetus for countries to develop requisite PCHL support services beyond sole investment in schools for the deaf.

HOW FAR CAN PCHL BE PREVENTED?
PCHL is an aetiologically heterogeneous condition almost equally attributable to genetic and environmental causes. The major contributors to neonatal mortality such as prematurity, birth asphyxia, sepsis and meningitis which have attracted substantial global health attention are also associated with PCHL in many survivors (table 1). Prevention of PCHL unquestionably should be preferred to later treatment

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### Table 1 Aetiological and risk factors for congenital and early-onset hearing loss

<table>
<thead>
<tr>
<th>Conditions</th>
<th>Primary Prevention Options</th>
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<tbody>
<tr>
<td><strong>Known Factors (40%-62%)</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Prenatal</strong></td>
<td></td>
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<tr>
<td>Family history of deafness</td>
<td>None</td>
</tr>
<tr>
<td>Consanguinity</td>
<td>Parental counselling</td>
</tr>
<tr>
<td><strong>Genetic: Syndromic</strong></td>
<td></td>
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<tr>
<td>Treacher-Collins syndrome</td>
<td>Genetic evaluation and counselling</td>
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<tr>
<td>Pendred syndrome</td>
<td>Discourage consanguineous marriages</td>
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<tr>
<td>Usher syndrome</td>
<td>As above</td>
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<tr>
<td>Waardenburg syndrome</td>
<td>As above</td>
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<tr>
<td>Jervell and Lange-Neilsen syndrome</td>
<td>As above</td>
</tr>
<tr>
<td>Others (eg. Hunter syndrome)</td>
<td>As above</td>
</tr>
<tr>
<td><strong>Genetic: Non-Syndromic</strong></td>
<td></td>
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<tr>
<td>Connexin 26 (35del G mutation)</td>
<td>Genetic evaluation and counselling</td>
</tr>
<tr>
<td>Connexin 31</td>
<td>Discourage consanguineous marriages</td>
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<tr>
<td>Others</td>
<td>As above</td>
</tr>
<tr>
<td><strong>Chromosomal</strong></td>
<td></td>
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<tr>
<td>Down syndrome</td>
<td>Genetic evaluation</td>
</tr>
<tr>
<td>Edward syndrome</td>
<td>Prenatal diagnosis and termination of pregnancy</td>
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<tr>
<td>Patau syndrome</td>
<td>As above</td>
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<tr>
<td>Turner syndrome</td>
<td>As above</td>
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<tr>
<td><strong>Acquired Congenital Infections</strong></td>
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<tr>
<td>Rubella</td>
<td>Rubella vaccination for girls and infants</td>
</tr>
<tr>
<td>Cytomegalovirus</td>
<td>Health education on secondary obstetric and childcare practices</td>
</tr>
<tr>
<td>Herpes Simplex</td>
<td>Prevention</td>
</tr>
<tr>
<td>Toxoplasmosis</td>
<td>Health education/caesarean section</td>
</tr>
<tr>
<td>Syphilis</td>
<td>Health education/treat mother</td>
</tr>
<tr>
<td><strong>Natal</strong></td>
<td></td>
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<tr>
<td>Lack of skilled birth attendant</td>
<td>Maternal education and improved obstetric and childcare practices</td>
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<tr>
<td>Mode of delivery</td>
<td>As above</td>
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<tr>
<td>Maternal hypertensive disorders in</td>
<td>As above</td>
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<tr>
<td>pregnancy</td>
<td>As above</td>
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<tr>
<td>Birth trauma</td>
<td>As above</td>
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<tr>
<td>Birth asphyxia</td>
<td>As above</td>
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<tr>
<td>Prematurity/Low birthweight</td>
<td>As above</td>
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<tr>
<td><strong>Postnatal</strong></td>
<td></td>
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<tr>
<td>Neonatal sepsis</td>
<td>Improved obstetric and neonatal care</td>
</tr>
<tr>
<td>Neonatal meningitis</td>
<td>Vaccination (especially in at-risk population)</td>
</tr>
<tr>
<td>Neonatal jaundice</td>
<td>Early detection and prompt treatment</td>
</tr>
<tr>
<td>Ototoxicity (eg. aminoglycosides)</td>
<td>Avoidance/rational use of ototoxic drugs</td>
</tr>
<tr>
<td>Undernutrition</td>
<td>Early nutritional intervention</td>
</tr>
<tr>
<td><strong>Unknown Factors (38%-60%)</strong></td>
<td>None. Further research required.</td>
</tr>
</tbody>
</table>

*Adapted from Olusanya and Newton.*

**APPROPRAISOES TO NHSP IN RESOURCE-POOR COUNTRIES**

Worldwide promotion of NHSP has been considered an important and achievable goal. With NHSP now established as a standard of newborn care in developed countries, the issue for developing countries is not ‘whether’ but ‘how’ such service should be delivered. However, early detection of the vast majority of infants with PCHL would require access to all eligible babies at birth or shortly thereafter. Less than 60% of deliveries in developing countries occur in hospitals. Deliveries outside hospitals in Africa are as high as 95% in Ethiopia, 91% in Somalia and 65% in Nigeria; and in South Asia, 85% in Bangladesh and 82% in Nepal. Community-based hearing screening programmes or a combination of hospital and community-based programmes are therefore most appropriate for majority of these countries.

The sheer number of annual live births along with an acute shortage of requisite resources in many countries makes the more ideal universal NHSP an unattainable goal on a national scale. In fact, with the exception of HIV/AIDS screening, which is backed by substantial international global health support, national screening programmes for health conditions in children are rare. The choice between universal or targeted NHSP in a hospital or community setting will largely be determined on case-by-case basis depending on available resources. This decision will be influenced by efficiency considerations such as overall yield, cost per baby screened, cost per infant detected and follow-up effectiveness.

Of the 110 countries eligible for this review (see box 1 and supplementary material), various forms of NHSP projects have been documented in China, India, Pakistan, Thailand, Philippines, Iran, Jordan, Egypt, Tunisia, South Africa, Nigeria, Cote d’Ivoire, Colombia, Cuba, Ecuador and Guatemala. NHSP projects have not been reported in the vast majority of the countries although the absence of evidence in the literature is not necessarily evidence of absence. While the feasibility especially in resource-limited settings. However, a significant proportion of children have idiopathic PCHL. The aetiology is probably unknown in 38% to 60% of affected children. The prevailing socioeconomic conditions and weak healthcare systems in many countries make complete elimination or significant reductions of the known causes still unattainable including mortality-related adverse perinatal and postnatal conditions. Primary prevention of genetic or hereditary aetiologies such as consanguinity is also likely to have limited effectiveness in many cultural settings.

Complementary secondary prevention through early detection and management of PCHL therefore becomes imperative to provide safety nets for those who unavoidably will be hearing impaired. Moreover, NHSP offers a more effective platform to investigate the probable causes of PCHL among the vast heterogeneous populations in developing countries as current epidemiological data on PCHL is derived substantially from studies conducted among deaf school-aged children.
Global child health

of NHSP has been confirmed in all reporting countries, setting up effective follow-up and monitoring systems to ensure timely completion of the required stages up to confirmatory tests was the most prominent challenge. The screening tools were automated transient or distortion product otoacoustic emissions (TEOAE or DPOAE) and automated auditory brainstem response (AABR) used singly or in combination with provision for repeat testing. Behavioural or questionnaire-based tests were rarely reported.

An informal experts’ consultation on infant hearing screening under the auspices of WHO has provided some guidance for the development of local NHSP programmes. After examining several practical issues including ethics and cost effectiveness of screening, withholding NHSP could not be justified for any population. However, the need to develop context-specific strategies based on experiences from pilot studies was emphasised. It was recognised that cost-effectiveness analysis in settings where healthcare services are financed substantially as out-of-pocket expenses will differ considerably from the practice in countries with free/publicly-funded national healthcare services.

Considering the constraints that prevent public health sectors from delivering requisite services in resource-poor countries, public–private partnerships may be required for a stepwise introduction of NHSP and related services. The role of governments and the global health community may be prioritised towards facilitating capacity building in human resource development, public education and instituting regulatory framework for best practices. Specific approaches to curtailing the burden of childhood hearing loss based on a country’s income profile have also been proposed. As with many public healthcare decisions, government’s involvement in the direct provision of NHSP-related services may be influenced by political, ethical and equity expediency rather than cost-effectiveness considerations. Effective advocacy by parent support groups, professional associations and the media may constitute a major catalytic force for this purpose.

SUPPORTING INFANTS DETECTED AS HAVING PCHL

The benefits of early detection of PCHL are best optimised with timely initiation of requisite (re)habilitative services particularly for satisfactory speech and language outcomes. The choice of appropriate communication and educational modalities for children with PCHL is still a subject of debate between the deaf and the hearing communities. However, the vast majority of children with PCHL have hearing parents who look forward to establishing auditory–verbal communication with their children. It is therefore essential that such parents are helped to achieve this goal as far as possible, failing which other communication methods can be instituted. Notwithstanding the superior advantages from auditory–verbal intervention, parents and health professionals need to know that even early introduction of sign language in the early months of life can provide parents and their child with a viable communication mode.

While the opportunity for literacy skills and education is limited it is preferred to outright neglect or the common practice where affected children are forced to beg for alms as a vocation in some communities. Intervention can also be targeted separately at achieving improved outcomes in reading and literacy skills while optimising overall educational achievement with a specific language base. It may also seek to establish appropriate family understanding and acceptance of hearing loss, reduce family stress as the child develops and improve social and emotional development throughout the school years. Educating parents with the aim of curtailing the incidence of child abuse and neglect and acting to promote and preserve the rights of children with disabilities are also worthwhile intervention goals.

To address the disincentive of high costs, there are increasing international efforts to make hearing aids more affordable and widely accessible in resource-poor countries. For example, prices as low as US$10 for body-worn analogue hearing aids and US$50 for digital hearing aids have been reported in India. While amplification devices may aid speech and language acquisition balancing the trade-offs between low cost and functionality for effective speech discrimination remains a challenge. Hearing aids are visible rehabilitative/assistive devices for a chronic condition rather than a one-off treatment like conventional medication and cannot eliminate all the consequences associated with PCHL. Even when these devices are provided at no charge, some parents may still be bothered by the associated stigma resulting in the devices being used less frequently thus compromising their effectiveness. Supporting parents to have realistic expectations with this intervention is therefore essential.

Cochlear implants may be more effective for some infants with severe-to-profound PCHL. However, the high cost also remains a major impediment towards their widespread use. Nonetheless, parents must be adequately informed of this possibility without undue prejudice to their perceived financial status. Efforts to promote this intervention must also be backed by requisite postimplantation support services to optimise speech and language outcomes.

ROLE OF PAEDIATRICIANS IN THE MANAGEMENT OF PCHL

Saving lives is a priority task for all paediatricians by virtue of their training and the overwhelming workload of routine clinical practice. However, as far as practicable the duty of care in routine newborn examination prior to discharge challenges paediatricians not to declare newborns with known risk factors but undetected PCHL as fit for discharge or without risk of subsequent developmental disability. For this reason, NHSP must be seen as a component of neonatal examination to facilitate prompt referral of infants requiring further evaluation to audiologists or otolaryngologists. Effective long-term management will require the services of (re)habilitation specialists including developmental paediatricians where available. Even where universal NHSP is impracticable immediately, timely, prompt referral of high-risk infants should be routine practice. Sudden discovery of PCHL in a child that had been previously declared ‘healthy’ undermines parental trust and confidence in the doctor. This may be construed as professional malpractice particularly in the presence of commonly reported risk factors such as consanguinity, admission for intensive or special care at birth and severe hyperbilirubinemia. Paediatricians are well placed to ensure that current maternal, newborn and child health (MNCH) services incorporate parental education on PCHL and the benefit of early detection. Local paediatric associations must demonstrate leadership and commitment for this vision through position statements. For example, the current leadership of the Indian Academy of Paediatrics has made NHSP a priority inspired by findings from local pilot projects.

Where basic, automated and simple-to-use hearing screening instruments such as TEOAE/DPOAE and AABR are not immediately available in individual hospitals, centralised screening centres serving several hospitals in a
community may be considered. Paediatricians can also participate in the design of evaluation studies to more accurately establish context-specific evidence on the impact of early detection and intervention on the quality of life of infants with PCHL. Given that a high proportion of deliveries occur outside hospitals, partnership with public health professionals is necessary for achieving a wider community impact.

CONCLUSIONS

The vast majority of infants with PCHL live in resource-poor countries where no form of hearing screening is offered routinely. Failure to identify and provide appropriate support for such infants in early childhood is likely to set them on a trajectory towards poor cognitive, linguistic and psychosocial development that irretrievably undermines educational and vocational attainment. Moreover, experience-dependent synaptogenesis of the developing human brain clearly supports priority attention for all infants at risk of suboptimal auditory development in the first year of life. While the opportunity cost of a lack of NHSP in resource-poor countries is multifaceted and substantial extending well beyond speech and language outcomes, paediatricians in these settings are uniquely positioned to promote NHSP within MNCH framework to facilitate timely intervention for infants with PCHL. Emerging global health interest in ECD now provides a unique opportunity for constructive engagement by individual countries and stakeholders to curtail the burden of this lifelong condition.

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