The Burden and Management of Infants with Life-long and Irreversible Hearing Impairment in Nigeria

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ABSTRACT

If left untreated or treated late and inappropriately, sensorineural hearing impairment has several adverse, irreversible and life-long health, psycho-social and economic consequences for the affected infants and their families. This seminar paper sets out to describe the burden, current practices and management options for sensorineural or permanent hearing impairment in early infancy for primary care physicians in Nigeria against the backdrop of their role as primary or first contact for all health consultations. Available studies suggest that up to 2.7% or 162,000 of the 6 million infants born annually may have permanent hearing impairment. The underlying aetiological factors may not be determined in the majority of the infants thus limiting the effectiveness of any primary prevention initiatives. However, the affected infants can be detected accurately with objective screening technologies such as otoacoustic emissions and auditory brainstem response in hospital or community settings. The ethical and scientific rationale for this intervention has been well established. Barring the challenge of parental follow-up default which is not insurmountable, affected infants and their families can be supported to establish appropriate auditory-based communication, avoid potentially harmful traditional therapies and child neglect commonly associated with childhood hearing impairment. Primary care providers in private and public practice have a crucial role in guiding parents to seek timely and appropriate services from ear care providers and child development specialists to ensure optimal child growth and developmental outcomes. On-going parental commitment to and active participation in the selected intervention programmes are essential to satisfactory long-term outcomes.

KEYWORDS: Early detection; infant hearing screening; targeted screening; auditory-verbal communication; sign language; rehabilitation; Africa; developing country.

INTRODUCTION

Hearing impairment is the most prevalent sensory disability in humans affecting all age-groups [1-3]. For example, of the 120 million babies born yearly in the developing world, about 718,000 are likely to have permanent or sensorineural hearing impairment at an estimated incidence of 6 per 1000 live births compared to 2-4 per 1000 for the developed world [4,5]. Sensorineural hearing impairment that is present at birth (congenital) or in the first 28 days of life (early-onset) has substantial short and long-term consequences across key developmental domains of motor, speech and language, cognitive and psychosocial skills in early childhood as well as subsequent educational and vocational attainment [6-9]. A third or more of infants with hearing impairment are also known to have additional neurodevelopmental disabilities [10], with Sub-Saharan Africa and Southeast Asia accounting for a disproportionate burden of developmental disabilities globally [11]. Of all developmental disabilities originating at birth or shortly thereafter, hearing impairment is of special interest because it is the most invisible yet detectable by current screening technologies and only begins to manifest through speech and language delay at a time (typically after 2 years) when the best of intervention can only but be sub-optimal [8,11].

As far back as 1995, the World Health Assembly passed a resolution urging all countries to take appropriate steps towards the prevention and control of major causes of avoidable hearing loss and for early detection in babies, toddlers and children within the framework of primary health care [12]. In the last decade, the early identification of hearing impairment has emerged as an essential component of neonatal care in the developed world [13] and to a limited but growing extent in some developing countries such as Brazil, Oman, India and South Africa [14-17]. This has been spurred by the availability of objective, simply-to-use and rapid hearing screening tests and the substantial scientific evidence on the benefits of early detection and intervention in the first year of life [8,9,13,18]. In fact, infants who are detected late (after 6-12 months) are associated with significant life-long deficits in speech and language as well as cognitive and psychosocial development which in turn severely restricts educational and vocational attainment [8,9]. More recently, a

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landmark report from the World Health Organisation reflecting contributions from Nigeria has outlined guiding principles for the implementation of newborn and infant hearing screening programmes based on a consensus position of experts from all regions of the world [19].

Since primary care physicians in both private and public sectors are usually the first and primary contacts for all health-related consultations, this seminar paper attempts to provide an up-to-date overview of childhood hearing impairment in Nigeria within the context of primary health care to facilitate the effective management of this irreversible and life-long disorder from birth or early infancy at all levels of health care delivery.

HEARING MECHANISM AND TYPES OF HEARING IMPAIRMENT

The peripheral auditory system consists of the external ear, the middle ear; and the internal ear (Figure 1). The process of hearing begins with the collection of sounds in the outer ear which are then transmitted through the middle ear into the inner ear. The tympanic membrane vibrates in response to incoming sound pressure waves and transmits energy through the ossicles or ossicular chain into the cochlear. Inside the cochlear are highly specialised hair cells which convert the incoming sound energy into electrical signals or impulses for transmission through the eighth nerve to the brain for processing and interpretation. Hearing is impaired if there is any obstruction or malfunctioning along this entire auditory pathway. For example, when the incoming sounds cannot be transmitted beyond the middle ear, a conductive hearing impairment is said to have occurred. Common causes of conductive hearing impairment include structural birth defects of the outer and middle ear, excessive build-up of cerumen, perforation of the ear drum and otitis media. With the exception of hearing impairment associated with structural defects or chronic otitis media, conductive hearing impairment is usually transient as it can be fully reversed after the underlying cause(s) have been treated. Hearing impairment is sensorineural or permanent when sound transmission is terminated in the cochlea or around the eighth nerve. This occurs when the specialised hair cells are damaged as a result of prenatal and birth related complications, viral and bacterial infections, (mis)use of ototoxic drugs such as antimalarials and antimicrobials especially aminoglycosides, as well as hereditary factors such as consanguinity or family history of deafness. When hearing impairment has both conductive and sensorineural components it is referred to as mixed. Sensorineural hearing impairment is the commonest auditory disorder in newborns or young infants (3 months old or less) and it is the primary focus of this seminar paper. Hearing impairment can be unilateral, affecting one ear, or bilateral affecting both ears. Hearing impairment can also be classified by time of onset as either congenital (occurring at birth), acquired (occurring any time after birth), early-onset (occurring within the first 28 days of life), delayed-onset (manifesting after the primary causal event) or progressive (manifesting gradually over time usually with increasing severity). The degree of hearing impairment and its effects when left untreated are usually classified as slight, mild, moderate, severe and profound as shown in Table 1 [5].

Table 1. Common effects of untreated childhood hearing impairment*

Average Hearing Level and Degree of Hearing Impairment	Receptive Language	Expressive Language	Activity Limitation/ Participation Restriction
0 -15 dB HL: *Normal Hearing*	Detects all speech signals	Normal range	None
16 - 25 dB HL: *Slight*	Misses up to 10% of speech sounds (e.g. unvoiced consonants) especially in difficult listening situations	Mild dysfunction in language learning	- Inappropriate response to sound - Learning difficulties - Poor social interaction
26 - 40 dB HL: *Mild*	Misses 25 to 40% of speech especially in difficult listening situations	Mild language retardation and speech problems	- Inattention - Learning difficulties - Behaviour problems
41 - 55 dB HL: *Moderate*	Misses 50 to 75% of speech	Moderate language retardation and poor speech intelligibility	- Learning dysfunction - Significant social problems
56 - 70 dB HL: *Moderately Severe*	Misses 75 to 100% of speech	Severe language retardation and speech problems	- Severe learning dysfunction - Stigmatisation and possible social isolation
71 - 90 dB HL: *Severe*	Misses up to 100% of speech at conversational level	Severe speech problems and language retardation	- Severe learning dysfunction - Stigmatisation and significant social isolation
> 90 dB HL: *Profound*	Misses all loud speech sounds except vibrations.	Visual cues essential for communication	- Complete social isolation

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THE BURDEN OF INFANT HEARING IMPAIRMENT IN NIGERIA

For many years, the burden of childhood hearing impairment in Nigeria was characterized solely by findings from studies conducted in mainstream schools and special schools for the deaf as well as tertiary hospitals [20]. Perhaps the most representative of the studies among school-aged children conducted to date in Nigeria was the national survey by the National Ear Care Centre in 2001 [21]. The prevalence of permanent hearing impairment of moderate-to-severe (41dB-90dB) or profound (>90dB) degree was reported as 6.9% which increased to 13.4% when infants with mild (25dB-40dB) hearing impairment were included. The rates are consistent with findings from an earlier study of 8 randomly selected schools in Lagos where the prevalence rates were reported as 8.9% and 13.9% based on the two respective thresholds [22]. These rates are the highest reported so far from sub-Saharan Africa [23]. However, it is difficult to accurately ascertain the proportion of school-aged children with hearing impairment in mainstream schools that is congenital or of early-onset usually from parental reports. Moreover, infants with severe-to-profound hearing impairment originating in early infancy are unlikely to be in mainstream schools although this problem is not peculiar to Nigeria. It is only through a universal infant screening programme that a more accurate estimate of the prevalence of congenital and early-onset hearing impairment can be achieved [24].

To date, only three studies on infant hearing screening have been reported in Nigeria [25,26]. The first two studies were conducted between 2005 and 2008 in Lagos under the auspices of the University College London. The first study was a hospital-based universal screening of all surviving newborns delivered at the Lagos Island Maternity Hospital. Of the 4178 screened under this programme 117 (2.5%) were estimated to have hearing impairment based on the proportion (12 or 0.3%) that completed the diagnostic tests [25]. The screening protocol consisted of a first-stage test with transient-evoked otoacoustic emissions (TEOAE) and a second-stage test with automated auditory brainstem response (AABR) discussed in greater detail in later sections of this article. The second study was a community-based universal screening of all infants below the age of 3 months who presented for BCG immunisation at four primary care centres in Lagos Island Local Government Area. Of the 7179 screened, 215 (3.0%) were estimated to have permanent hearing impairment, based on the 71 (1.0%) who completed the diagnostic assessment. Overall, of the 11,897 infants tested, between 83 (0.7%) and 332 (2.7%) infants had permanent hearing impairment. In the last study by Okhakhu et al conducted in Benin City over a 3-month period, of the 400 neonates screened, 90 (22.5%) infants failed the hearing test either in one or both ears [26]. However, the screening protocol was limited to otoacoustic emissions and did not provide an adequate assessment of the hearing status of the infants. It was therefore likely that a significant proportion of those who failed the screening test would have passed the more objective and reliable auditory brainstem response test. Overall, the reported rates in Nigeria are the highest in sub-Saharan Africa and among the highest in the developing world [27].

While it is difficult to extrapolate the findings from the three studies from Southern Nigeria to the entire country, the evidence from the national survey by the National Ear Care Centre would suggest that the comparable rates in Northern Nigeria are likely to be higher [21]. The factors likely to account for such a pattern include the potential contribution of meningitis and consanguinity which are more common in that part of the country. Barring the limitations of available studies, an estimated 42,000 and 162,000 of the 6 million infants born annually in Nigeria are likely to have permanent hearing impairment with life-long consequences. This should be of public health concern even before more robust estimates from population-based prospective studies sufficiently representative of the six geopolitical zones in the country become available.

AETIOLOGY AND RISK FACTORS FOR INFANT HEARING IMPAIRMENT IN NIGERIA

Several studies have explored the aetiology of childhood hearing loss in Nigeria but are predominantly hospital-based involving convenient samples or among children in schools for the deaf [28-32]. The proportion of infants in these samples was often quite limited reflecting the delays in presenting to hospitals for treatment. The most up-to-date overview of such studies perhaps was published by Dunmade et al in which the unknown category reported across studies varied from 36.2% to 59.7% [28]. Notwithstanding the limitations in the clinical diagnosis of aetiological factors in available studies, genetic factors such as Usher's and Waardenburg's syndromes as well as environmental factors such as congenital rubella syndrome, birth asphyxia, hyperbilirubinaemia and ototoxic drugs especially gentamycin have been documented [28-32].

Infant hearing screening programmes provide the most representative samples for aetiological investigation in early infancy. However, a typical diagnostic protocol consists of several advanced tests including genetic evaluation for DNA isolation, MRI, Karyotyping and perchlorate tests that are not routinely offered in a developing country like Nigeria. Even where these tests are available, the aetiology of congenital hearing impairment may not be known in almost half of the affected infants. An alternative and common approach is to conduct a retrospective analysis of the distribution of known aetiological or risk factors among infants

detected with hearing impairment. For example, the commonest risk factor among the 98 infants who failed the two-stage screening with TEOAE and AABR in the hospital-based universal newborn hearing screening programme in Lagos were undernutrition (34.7%), birth asphyxia as indexed by 5-minute Apgar score <7 (31.6%) and admission into intensive/special care unit (19.4%). None of the known risk factors could be identified in 48% of the infants. In contrast, under the community-based programme it was not possible to ascertain the clinical risk factors found in the hospital. However, among the 128 infants who failed the hearing tests 58.6% were undernourished while 49.2% were delivered without skilled attendants at birth. Some 20% of the infants had no identifiable risk factor. About 10% of the infants in each of the two programmes were treated for neonatal jaundice while family history of deafness was not reported by any mother. Additional evidence (unpublished) suggests that microcephalic infants in Nigeria are also at risk of hearing impairment.
The large proportion of infants with idiopathic hearing impairment in Nigeria like in many other developing countries makes it imperative to implement programmes for the early detection of infants who are unlikely to benefit from any primary prevention programmes directed towards curtailing the known risk factors. Once identified through effective systematic screening programmes, the affected infants must be provided with appropriate support to reduce the burden of the disability in all crucial domains of early childhood development.

EARLY DETECTION OF INFANTS WITH HEARING IMPAIRMENT

CURRENT PRACTICES TOWARDS INFANTS WITH HEARING IMPAIRMENT

There is currently no form of routine or systematic screening for hearing impairment at any level of healthcare delivery in Nigeria. As in the developed world, parents are often the first to suspect the existence of a hearing loss as a result of a child's inattention, erratic response to sound or speech delays. This was evident in a recent study conducted in the two largest public schools for the deaf in Lagos with a total enrolment of 429 pupils (mean age: 10.3 years) [33]. 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 six 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 least associated with hearing difficulty by parents (1%).

Routine hearing screening offers parents of a hearing-impaired child the knowledge of the special needs of their apparently normal baby as early as possible. Thus it helps to avoid the "diagnostic odyssey" of trying to unravel a suspected disorder. For example, in the absence of screening, hearing impairment is unlikely to be detected until the parents or caregivers observe a child's inability to respond to sound, inappropriate behaviours or speech and language defects when compared to their peers as from 12-18 months of age. During this process, suspecting parents are often anxious, confused, and make false assumptions about the nature, degree and full effects of the condition until they receive appropriate professional attention. And this process may often entail making consultations with a variety of both orthodox and traditional "service providers" who have no clue about the condition of the child. Parents and their infants with hearing impairment certainly have a lot to benefit from improved management of this condition.

OVERVIEW OF INFANT HEARING SCREENING

"Screening" can be defined as the systematic application of a test or enquiry to identify individuals at sufficient risk of a specific disorder that will benefit from further investigation or direct preventive action, among people who have not sought medical attention because of symptoms of that disorder [34]. When applied to hearing impairment, the process of screening should identify infants with hearing impairment for whom further action is warranted (test-positives) and infants without hearing impairment for whom no further action is warranted (test-negatives). It is highly unlikely that any hearing screening test can accurately distinguish all infants with hearing impairment from those without due to the inherent differences in biomedical investigation and test algorithms. Consequently, a hearing screening test usually results in four main outcomes:

A. Infants with hearing impairment accurately identified (True-Positives)
B. Infants without hearing impairment accurately identified (True-Negatives)
C. Infants with hearing impairment not accurately identified and classified as having normal hearing (False-Negatives)
D. Infants without hearing impairment not accurately identified and classified as having abnormal hearing (False-Positives)

The performance of an infant hearing screening test is therefore evaluated on the basis of the following parameters:

- Sensitivity probability of a positive test in infants with hearing impairment or the percentage of infants with hearing impairment correctly detected.
- Specificity probability of a negative test in infants without hearing impairment or the percentage of children without hearing impairment correctly detected as having normal hearing.
- False Positive Rate (FPR) probability of an infant without hearing impairment testing positive or the percentage of infants without hearing impairment

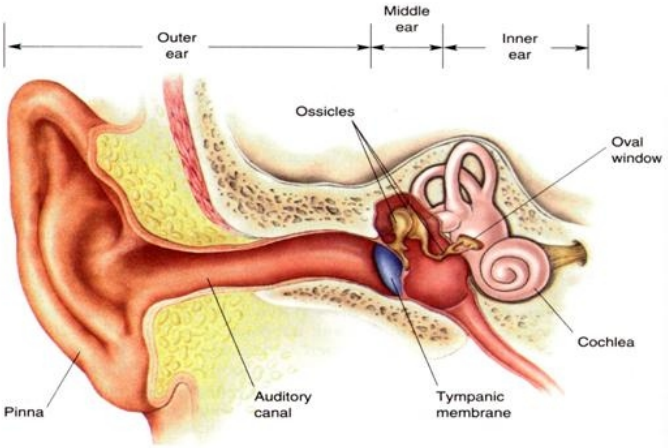

Figure 1. The Peripheral Auditory System

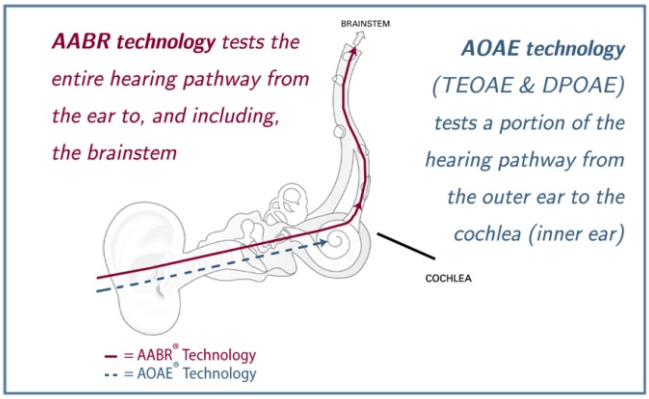

Figure 4. Pathways for Hearing Screening Tests
[http://www.natus.com/products/hearing_screen/echo.html]

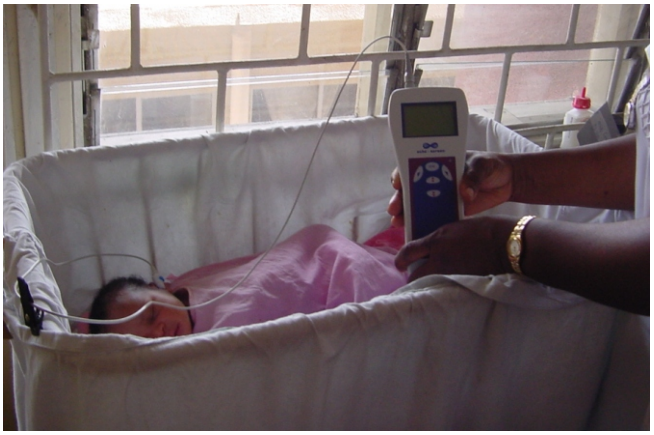

Figure 2. An automated otoacoustic emissions screener in use at a Lagos hospital

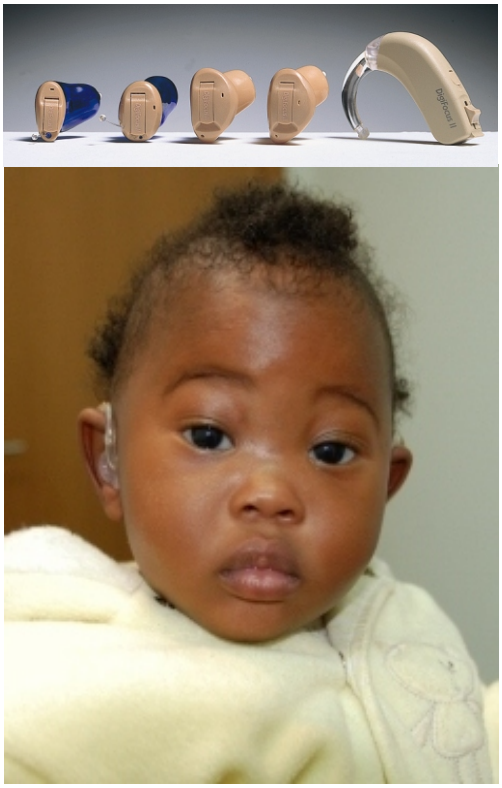

Figure 5. Infant fitted with hearing aids

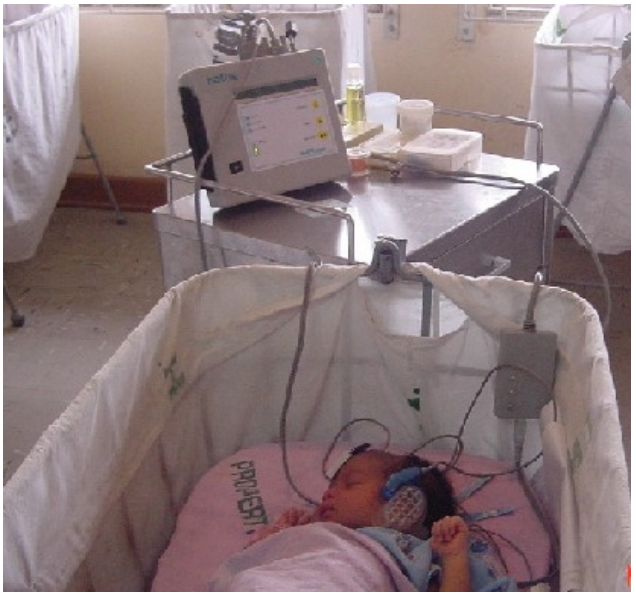

Figure 3. An automated auditory brainstem response screener in use at a Lagos hospital

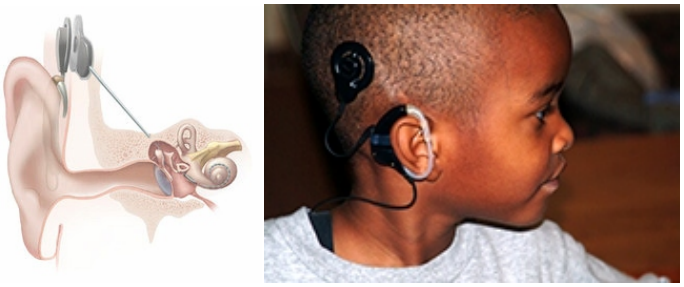

Figure 6. Child fitted with cochlear implant

who had positive test results.

- Positive Predictive Value (PPV) probability of an infant having hearing impairment when the test is positive or the percentage of those with positive test results who actually have hearing impairment.

Whilst screening has the potential to improve quality of life through early diagnosis, it cannot offer a guarantee of protection against developing the full-blown condition. It is most useful primarily to curtail the burden or consequences of the condition. In practice, an ideal hearing screening test would be simple to apply, safe, reliable and valid. It is reliable if it provides consistent results and valid if it detects the majority of infants with hearing impairment (high sensitivity); does not pick most infants without hearing impairment as failing the test (high specificity) or the percentage of infants without hearing impairment among those with positive test results is very low (low FPR); and if the percentage of those with hearing impairment among those with positive test results is high (high PPV).

TOOLS FOR HEARING SCREENING

All hearing tests measure an individual's level of response to sound. Current techniques for hearing screening in all age groups can be classified into objective and subjective (or behavioural) tests. Objective tests are those that do not require the patient to respond physically to sound or auditory stimuli such as automated otoacoustic emissions (OAE) and auditory brainstem response (AABR). In contrast, subjective tests such as pure-tone audiometry (PTA) or visual response audiometry (VRA) require a visible response by the patient. Subjective tests are not suitable for screening babies and very young infants but are useful as confirmatory tests in older children.

OTOACOUSTIC EMISSIONS (OAE)

Otoacoustic emissions are low intensity sounds generated from the outer hair cells of the cochlea in response to audible sounds. There are two main types of OAEs namely, transient-evoked otoacoustic emissions (TEOAE) and distortion-product otoacoustic emissions (DPOAE). OAE is a physiological test for the specific measure of the integrity of the outer hair cells in the cochlea**.** OAE, also known as cochlear echoes, are low intensity sounds originating from the active amplification of the outer hair cells and can be elicited in response to clicks or tone bursts presented to the ear through a light weight probe that houses both a transducer and microphone/receiver (Figure 2). The emissions are then matched with a built-in template for normal values to determine a 'pass' or 'fail' result. The test is relatively quick, non-invasive and does not require sleep or sedation. The recording often takes seconds and can be administered without audiological expertise. However, the test is also sensitive to excessive internal noise from patient or ambient noise in the test environment and will not detect any retro-cochlear dysfunction of the inner hair cells and beyond such as auditory neuropathy spectrum disorder (ANSD). This disorder is a type of hearing impairment in which normal outer ear cell function of the cochlea co-exists with abnormal or dys-synchronous auditory brainstem response. A typical OAE instrument is light, portable and powered by an-inbuilt rechargeable battery that can last many hours of continuous use.

AUTOMATED AUDITORY BRAINSTEM RESPONSE (AABR)

The ABR is an electro-physiological measure of the function of the auditory pathway from the eighth cranial nerve through the brainstem. The major advantage of this test which is an electrical recording from three surface scalp electrodes to auditory stimuli is the fact that it is not state-dependent as recordings can be obtained when babies are sleeping or sedated. In addition, the response is significantly correlated with the degree of hearing loss. In general, the click-evoked threshold predicts behavioural audiometric threshold in the 1,000 to 4,000 Hz range within 10 to 15 dB HL. It is therefore valuable as a confirmatory test in infants. The automated version of ABR (AABR) is designed for screening purposes (Figure 3). A typical AABR instrument is also powered by an-inbuilt rechargeable battery and when activated delivers at least one thousand soft-click stimuli at 35 dBnHL to the newborn's ears through disposable flexi coupler earphones at a rate of 37 clicks per second. The responses to the auditory stimuli are recorded with three surface jelly tab sensors or electrodes placed over the vertex, nape and the shoulder or the cheek. A “pass” or “refer” is displayed based on the manufacturer's internally programmed template-matching algorithm for the measured ongoing brain wave or auditory brain stem response. However, it may be difficult to conduct this test without sedation in infants older than three months due to restlessness during testing.

CONDUCTING A LOCAL INFANT HEARING SCREENING PROGRAMME

Screening with OAE and AABR have been demonstrated to be feasible in Nigeria in both hospital and community settings [26,35,36]. The technologies can be used by nurses or primary care workers after few hours of training. There is no hard and fast rule about the choice of technology or the protocol to be used. However, a two-stage screening with an initial OAE followed by AABR for all OAE referrals has several advantages over a single or two-stage screening protocol with either OAE or AABR. For example, while OAE is commonly preferred for the initial screening, the referrals can be excessive especially in busy hospitals where the discharge policy is less than 48 hours. Introducing AABR will reduce the pre-discharge referral rates substantially thus minimising the burden

on follow-up services. The combination of OAE and AABR provides the opportunity to evaluate the entire auditory pathway as shown in Figure 4 and thereby facilitates the identification of infants with auditory neuropathy. No screening protocol is perfect. It is therefore essential to set up a surveillance scheme that will facilitate the detection of infants who will be unavoidably missed by the choice of a particular screening strategy. While universal screening of all eligible infants is ideal, resource-constraints would demand that priority be given to high-risk infants. These will include for example, infants born outside hospitals without skilled birth attendants, those with a history of severe birth asphyxia, severe neonatal jaundice, admission into NICU/SCBU or who exhibit signs of early malnutrition [37].

Whatever the screening method used, consent must be sought from at least one of the parents or legal guardians before a screening test if offered. Sufficient time must be given to allow the patient to make a decision without feeling coaxed or intimidated especially in helping them to understand the consequences of declining screening for their babies [38]. The most opportune time to begin discussing the importance of screening with parents, therefore, is probably during maternal and child health clinics, particularly before delivery. The arrival of the newborn is a joyous and emotional event for the family and the disclosure of a permanent abnormality in an apparently normal baby must be handled with sensitivity. Parental reaction to this information would normally be characterised by shock, denial, grief and depression [39]. However, evidence from the studies conducted so far shows that parental consent is readily given especially if screening is presented within the context of the routine neonatal examinations, which parents expect shortly after delivery or before hospital discharge. Parents are also likely to accept current hearing screening tests because they are, painless, non-invasive and quick to administer. Many parents would prefer not to confront the future guilt and regret for failing to provide the best possible help for their child within their control for an invisible condition that will inevitably become obvious.

Overall, the screening procedure must be supervised and monitored by ear care specialists such as audiologists, otolaryngologists or developmental paediatricians with relevant expertise in audiology/audiological medicine. Any infant hearing screening programme especially in a hospital-setting ideally should be managed by paediatricians within the context of the overall developmental trajectory of infants detected with hearing impairment with or without other medical conditions requiring specialist attention. It is an essential component of modern routine newborn examination prior to hospital discharge and should ideally be complemented with blood spot tests for the early detection of birth defects such as sickle cell, phenylketonuria (PKU) and hypothyroidism as far as practicable. Given the possibility of false-positive or false-negative outcome, it is important to present the result of any screening test as an indication for further confirmatory tests rather than as an evidence of impairment.

CONFIRMATION OF HEARING IMPAIRMENT

Infants who fail the hearing screening tests should be referred for diagnostic evaluation with audiological and medical components provided and supervised by ear care and medical specialists [14]. The audiological evaluation typically will include a medical and family history, tympanometry with high frequency (1000 Hz) probe tone for infants less than 4 months old; frequency-specific assessment of auditory brainstem response; click-evoked auditory brainstem response with insert ear phones; and/or visual reinforcement audiometry for infants older than 6 months. The medical component, often prompted by the outcome from the audiological evaluation, will include comprehensive clinical history, family history of deafness, physical examination as well as radiological and laboratory investigations to facilitate possible determination of the aetiology of hearing impairment where indicated. Urine culture for detection of cytomegalovirus may also be considered.

A major challenge common in many countries is failure by some mothers to present their infants who failed the screening tests for diagnostic evaluation. For example, of the 98 infants who failed the screening tests and were referred for diagnostic evaluation under the first hospital-based programme in Nigeria, 85 (86.7%) did not return compared to 30.5% (39/128) under the community-based programme even though all services under both programmes including the provision of hearing aids where required were offered at no charge to parents [25]. Several socio-cultural factors appeared to have contributed to the poor return rates in this population. Firstly, unfavourable attitudes and superstitious beliefs towards childhood deafness and other disabilities could be a major disincentive for follow-up compliance. For instance, among the predominant Yoruba ethnic group in Lagos, having a child with hearing impairment is perceived as a curse, a spiritual attack or divine punishment from a deity which is a source of stigma and shame for the affected family [40]. This attitude had minimal effects during the first-stage screening because mothers were already at the screening venue or because all infants regardless of their hearing status were screened and the vast majority usually would pass the tests. Secondly, despite assurances given to mothers when communicating screening results and the uncertainty regarding the final diagnostic outcome, mere failing the screening test sets in motion anxiety about the possibility of a hearing impairment and the associated consequences in an

apparently normal child. Some mothers were afraid or reluctant to face this thought at such an early age. Even when mothers promised to return they did not. Thirdly, the perception of hearing loss as non-life-threatening in an environment characterised by an overwhelming attention to child survival and little interest on developmental prospects of survivors is also a potential barrier to satisfactory return rates. It was also possible that mothers may not feel sufficiently motivated to return for follow-up if the communication from their trusted health professionals did not convey the seriousness of the a potential hearing impairment which is likely to be the case if healthcare providers are not themselves persuaded that permanent hearing impairment is a serious health condition. It was not uncommon for both mothers and their healthcare providers to hold the view that a health condition that does not kill does not hurt [41].

OPTIONS FOR MANAGING INFANTS DIAGNOSED WITH HEARING IMPAIRMENT

Permanent hearing impairment adversely affects all crucial developmental domains of speech and language, motor, cognition and psychosocial wellbeing as well as literacy skills, education and vocational attainment. But the effects of hearing impairment and the developmental needs of affected children are varied and distinct from child to child. Upon confirmation of hearing impairment healthcare providers are expected to guide parents on the resources and options available for supporting the affected infants and their families. The scope and goals of any intervention must be matched with the specific needs of the child (and the parents). The primary goal and rationale for early hearing detection worldwide is to facilitate communication between infants with hearing impairment and their parents firstly, and with those they have to interact with as they grow. Although the range of services required for the effective management of hearing impairment transverses both medical and non-medical disciplines such as paediatrics, otolaryngology, audiology, speech and language therapy, psychology, education and social services, it is essential for every medical personnel to have an overview of the special needs of the affected children to enable them offer informed counsel and guidance to parents if consulted. The major communication options for infants with hearing impairment can be broadly classified as auditory and non auditory-based modes.

AUDITORY-BASED COMMUNICATION

Any communication approach geared towards the development of spoken language is referred to as "auditory-based". It entails the use of hearing devices to facilitate full integration of the child into mainstream society including educational system. The two main hearing devices available to infants with hearing impairment are hearing aids and cochlear implant. A hearing aid is an electronic device that consists of a microphone, amplifier and a receiver (Figure 5). Sound signals are received through the microphone and sent to the amplifier, where they are processed and amplified according to preselected settings. The amplified sounds are then transmitted via the receiver inside the device on to the user's (infant's) ear drum for subsequent processing through the inner-ear to the brainstem (Figure 1). The device is powered by lithium battery that is replaced periodically depending on the usage. A cochlear implant is also an electronic device placed under the skin behind the ear and can only be fitted by an otolaryngologist through surgical procedure. Externally, a cochlear implant consists of a microphone which picks up sound signals from the environment and a speech processor which selectively filters signals and sends them through a thin cable to a transmitter (Figure 6). The transmitter is a coil held in position by a magnet placed behind the external ear, and transmits power and the processed sound signals to the internal section of the device by electromagnetic induction. The internal component consists of a receiver and stimulator secured in bone beneath the skin, which converts the signals into electric impulses and sends them to the brain through the auditory nerve system.

Although all hearing aids amplify sounds, not all hearing aids are effective for the development of spoken language. Considerable expertise is required for the selection and fitting of appropriate hearing aids. Furthermore, effective auditory-based intervention requires more than the fitting of hearing devices [42]. It requires active, ongoing, culturally-appropriate collaboration between the service provider and the child's family within the context of the overall developmental status of the child after appropriate assessment by a paediatrician. Parents must be required to be active participants in the intervention programme and discouraged from shifting more attention to siblings without special needs at the expense of the child with hearing impairment. A full discussion of the prerequisites for optimal outcomes is outside the scope of this review. However, the service provider must be sensitive to parental emotions, stress and grief as they adjust to the special and often unfamiliar needs of an apparently healthy child. They must also be committed to employing evidence-based techniques to facilitate developmentally appropriate language skills and enhance the family's understanding of its infant's strengths and needs, build family support and confidence in parenting the child and promote the family's ability to advocate for the child especially for suitable educational placement.

NON AUDITORY-BASED COMMUNICATION

The most common and oldest form of intervention for children with hearing impairment in developing countries including Nigeria is enrolment in schools for deaf children where sign language is the predominant

mode of communication. In Nigeria, enrolment in the schools for the deaf is principally due to the considerable delays in detection and poor awareness among health professionals and parents. In one study, it was reported that doctors were most commonly consulted for help (77%), but that the majority of children (80%) were rarely provided with hearing aids because children were often considered too young to be fitted with these devices [33]. The primary mode of intervention often suggested to parents, was enrolment into a school for children that are deaf where sign language is the sole mode of communication [33]. Even then, only about 6% of the children were enrolled in the school by 6 years of age. Consequently, the reading and comprehension abilities of these children are about 50-60% of their chronological age by the time they leave school typically at the age of 18 years. From another study in Lagos it was observed that parents from lower socio-economic class were less likely to enrol their hearing-impaired children in the schools for the deaf [43]. This would suggest that majority of children with hearing impairment from lower socio-economic classes were unlikely to receive any form of intervention for communication and would be more susceptible to abuse and neglect [40,44]. In addition, the use of traditional and unorthodox therapies such as medicinal plants and animal fat for deafness with doubtful efficacy is a common recourse for Nigerian parents as in other developing countries because of ignorance and superstitious beliefs [45-47].

CHOICE OF COMMUNICATION MODES FOR INFANTS WITH HEARING IMPAIRMENT

The choice of appropriate communication and educational modalities for infants with hearing impairment has been a subject of debate among service providers [48-50]. Since the vast majority of parents of infants confirmed with hearing impairment use spoken language their primary interest is on how to establish verbal communication with their children as soon as possible. Optimal speech and language development have very strong linkages with most of the remaining skills and for that reason it has often been the primary focus of many early intervention programmes. Parents should therefore be supported to achieve this goal as far as practicable failing which other modes of communication could be instituted promptly in a manner that preserves parental autonomy.

The first hurdle under auditory-based approach is parental acceptance of hearing aids for their babies at such an early age because hearing aids are associated more commonly with elderly people. Hearing aids worn behind-the-ear publicly announce an invisible disability in a child, which could be culturally embarrassing to parents and cause them great stress in responding to inquiries from neighbours and friends about these "strange" devices in an apparently normal child. During the initial stages, some parents may be vulnerable to the not uncommon misconception that hearing aids are not necessary because the child will outgrow the disability. Next barrier is the cost of hearing aids and other accessories including ear moulds and batteries which have to be replaced at various stages. Many of these factors apply to an even greater extent to the use of cochlear implants and influence the parents' decision-making process. In Nigeria, the requisite expertise and resources for supporting infants fitted with cochlear implants are still lacking but can be developed over time. Until such support services can be assured this option should not be contemplated or offered to parents on ethical grounds. The recommended first line communication option for infants with hearing impairment is the use of hearing aids based on cost consideration and level of available support services.

It is not uncommon to find that even after making an informed and appropriate choice, parents still seek more time to adjust to the new reality and it is always prudent not to pressure them to immediately follow up on their choices. A great motivation for parents often comes from the assurance and evidence that all the efforts involved in supporting the child are worthwhile. Testimonials of other parents with successful experiences are valuable and just as powerful as the displeasure of unsatisfied parents to prospective parents, which is perhaps the greatest challenge for service providers in developing countries. Parents would make any necessary financial sacrifice in anticipation of visible progress in speech and language development; however, the challenge for the interventionist is securing the ongoing parental participation crucial to achieving satisfactory language outcome that is commensurate with the quality of the services provided.

Parents and health professionals need know that even early introduction of sign language in the early months of life still provides parents of deaf children with a communication mode and has significant benefits for the cognitive and psychosocial development of the child [42,51,52]. Notwithstanding its limitations, sign language offers an opportunity for literacy skills and education and is preferred to the common practice where deaf children are forced to beg for alms as a vocation. Early sign language training can be offered to parents and the child in the various schools for the deaf in the country. Presently, there are special schools for the deaf in majority of the 36 states in Nigeria owned/managed by government or missionary organisations. In addition, there are institutions that cater for children with special needs such as hearing loss and blindness across the country. Similarly, intervention to reduce child abuse and neglect can be achieved with better parental education and introduction of legislation against maltreatments. Vocational centres can also be established to expose children with hearing impairment to various trades to enhance their economic

independence.

Intervention can also be targeted independently at achieving results in the other areas [53]. For instance, early intervention targeting sensory and perceptual skill development may include but not restricted to the provision of amplification devices. Intervention goal for language development may focus on enhancing parent/infant communication in the chosen or most feasible communication modality and developing verbal and reasoning skills to support literacy attainment [51,54]. Similarly, intervention may simply focus on increasing reading and literacy skills as well as optimising overall educational achievement with a specific language base [55]; or intervention in the psychosocial domain may seek to establish appropriate family understanding and acceptance of hearing impairment, reduce family stress as the child develops and improve social and emotional development throughout the school years [53].

CONCLUSION

Every year up to 2.7% or 162,000 of the 6 million live births in Nigeria have permanent hearing impairment which is one the highest rates worldwide. The underlying causes in more than two-thirds of the affected infants cannot be determined thus foreclosing primary prevention. Moreover, current healthcare practices do not include routine hearing screening from birth thus resulting in considerable delays in detection and management. However, there is demonstrable evidence that infants with permanent hearing impairment in Nigeria can be detected early and more accurately with available modern and objective hearing screening technologies such as otoacoustic emissions and auditory brainstem response audiometry. So far most of the affected infants are enrolled in schools for the deaf where the predominant communication mode is sign language. However, opportunities exist for providing auditory-based intervention services to facilitate the development of spoken language preferred by the vast majority of parents. Primary care providers have a crucial role to play in guiding parents to make appropriate choices for achieving the goal of optimal developmental outcomes for their children with hearing impairment from early infancy.

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