Fighting Hearing Loss in Children - A Clarion Call

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Abstract: Hearing impairment has serious consequences in children. Language and speech development, communication, educational achievement, skills acquisition and emotional and social wellbeing are significantly hindered in the presence of hearing loss. Affected children face many obstacles since spoken language is the predominant medium of communication and social interaction. Adequate auditory stimulation, especially in early childhood, is the foundation for speech and language development as well as the acquisition of literacy and other skills. Hearing loss is a threat to the growth and development of children and the burden that it creates both for the children and the community at large, especially in developing countries, is huge and unacceptable. This article briefly reviewed the prevalence, burden, causes, consequences and the importance of testing to facilitate early diagnosis and rehabilitation of hearing loss in children. It is intended to sensitize family physicians, primary care workers, other health workers and caregivers of children to facilitate early recognition and referral of children with hearing loss and to advocate with policymakers to the importance of creating and supporting policies to prevent hearing loss in children and mitigate its effects on affected children and on the community.

Key words: Hearing Loss • Children • Burden • Early Detection • Rehabilitation

INTRODUCTION

Hearing impairment is the most prevalent disabling condition globally [1-4]. More alarming is the fact that seventy five percent of people with hearing loss are believed to live in developing countries [5]. According to the World Health Organization statistics, there were 120 million individuals with a disabling hearing loss globally in 1995. By 2005, this figure had doubled to 278 million. And according to 2011 estimates the number has increased to 360 million people - over 5% of the world’s population. If milder cases of hearing loss are included, almost 10% of the world population are affected by hearing loss [1-4]. Since the world population is constantly increasing, it is reasonable to expect that the number of people suffering from hearing loss will also increase if care is not taken. Hearing loss is very common in children and its effects can be devastating. The aim of this review is to sensitize health workers especially family physicians, primary care workers and other caregivers of children in order to facilitate early recognition and referral of children with hearing loss and to advocate with policymakers to the importance of creating and supporting policies to prevent hearing loss in children and mitigate its effects on affected children and on the community.

Prevalence and Burden of Hearing Loss in Children:
Population-based studies in Europe and North America have identified a consistent prevalence of approximately 0.1% of children having a hearing loss of more than 40 decibels (dB) through review of health or education records, or both. Other international studies using different methods or criteria (such as screenings, questionnaires and less severe decibel thresholds) have reported higher estimates [5]. About 32 million children are believed to suffer from disabling hearing loss according to 2011 WHO estimates [6]. And childhood hearing loss is estimated to constitute approximately 25% of the almost 10% of the world population believed to be affected by at least mild hearing loss [1, 2].

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Hearing loss is recognized as one of the most common birth defects and the number of children affected is increasing at an alarming rate [7]. International statistics for children with hearing impairment are reported to be 2-6/1000 live birth [8]. Significant hearing loss is present in 1 to 6 per 1000 newborns [9]. Every year in the United Kingdom, one in every 1000 children is born with a permanent hearing impairment [10]. In the United States of America, the prevalence of hearing loss among children aged three years or older was 13.2 million in 1971, 14.2 million in 1977, 20.3 million in 1991 and 24.2 million in 1993 [11, 12]. Congenital or early onset permanent bilateral hearing loss affects an estimated 798,000 newborns annually [13]. At least 90% of these reside in developing countries around the world [14]. This means that almost 2000 babies with hearing loss are born daily in developing world regions where they have no prospect of early detection [13]. Other figures that have been reported include 11.3% prevalence of minimal sensorineural hearing loss in school-age children in the USA by Bess et al. [15] and either low frequency or high frequency hearing loss in 14.9% of children of 6-19 years of age in the 3rd National Health and Nutrition Examination Survey reported by Niskar [16]. Also, Zakzouk and Al-Anazy [17] in Saudi Arabia gave the prevalence of hearing loss in age group 5–15 years as 13%.

In developing countries, infant hearing loss has been referred to as a silent epidemic [18] because despite its widespread prevalence, it is not detected by routine clinical examination and without appropriate screening, caregivers may not be aware. Also, being non-life-threatening, it does not receive the priority or visibility on global health care agendas that its long-term sequelae deserve [18] and 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 [19].

In Nigeria, from where we write, some studies have been conducted, though these were confined to specific age groups or certain geographical areas. The prevalence of hearing loss in school-aged population in Nigeria has been found to range from 6.7 to 8.9% in some studies [19]. It is also estimated that 6000–27,000 babies with permanent congenital and early-onset hearing loss (PCEHL) will be born in Nigeria annually and about 5000–22,000 of these will live beyond five years of age [20].

The impact of hearing loss in children is huge. Without early intervention these children are assigned to a life of deprived language development, restricted academic prospects, limited literacy and poor vocational outcomes [21]. Even with intervention, with improvements in life expectancy, childhood onset hearing loss has significant implications for long-term economic costs not only to individuals and families, but to communities and countries [1, 18, 21, 22]. It has therefore been suggested that the global burden of disease for childhood hearing loss may be significantly higher than for adult-onset hearing loss [21, 23] which currently ranks third on the global causes of years lived with disability (YLD) index and 15th on the disability adjusted life-years (DALY) index (one of four non-fatal conditions among the 20 leading contributors to the global burden of disease) [2, 24].

According to the Centers for Disease Control and Prevention [5], during the 1999 - 2000 school year, the total cost in the United States for special education programs for children with hearing loss was $652 million, or $11,006 per child and the lifetime educational cost was estimated at $115,600 per child [25]. It is expected that the lifetime costs for all people with hearing loss who were born in 2000 will total $2.1 billion [5]. The costs include direct medical costs (6%), direct nonmedical expenses, including special education (30%) and indirect costs, such as the value of lost wages (63%). They do not include other expenses, such as hospital outpatient visits, sign language interpreters and family out-of-pocket expenses [5]. Therefore, the actual economic costs will be even higher [5].

Aetiology and Onset: The precise aetiology or onset of hearing loss in children is often difficult to determine. Screening for early hearing detection and intervention is therefore imperative as a secondary prevention strategy for hearing loss in children [26]. Universal newborn hearing screening (UNHS) is promoted in developed countries as an early detection strategy for permanent congenital and early-onset hearing loss (PCEHL) [27]. Pilot programmes are already being implemented in Brazil, Mexico, South Africa, India and the Middle East. In Nigeria, there have been a few sporadic attempts in some hospitals, schools and communities [28]. However, routine screening for childhood hearing loss is still rare in most developing countries. The benefits and challenges of introducing UNS in developing countries have been recently articulated by Olusanya et al. [28].

Parental suspicion prompted by a child’s inappropriate or lack of response to sound is the primary mode of detecting children with PCEHL in Nigeria [29]. This passive detection usually occurs at a mean age of 22 months, which far exceeds the recommended early detection threshold of three months [30]. Parental
suspicion is commonly earlier for children with severe to profound PCEHL than those with mild to moderate PCEHL and even though it often helps to arouse a need for investigation, it is neither sufficient nor reliable.

It is important to test a child’s response to sound very early. Early discovery of any physical or intellectual condition that deviates from normal may lead to better and greater possibilities in management [31]. A child with even a mild hearing loss may suffer some degree of isolation during crucial stages of speech development [32]. Also, early medical management, enlightened early training to parents in language skills and placement in an appropriate educational setting (special schools or other facilities where enriched speech stimulation, speech reading instruction, auditory training and training in the use of hearing aids can be provided) may make it possible for the child with hearing impairment to fit into a regular classroom [33].

**Classification of Hearing Loss**

**Degree of Loss:** Sound should normally be perceived at a threshold that lies between 0 and 25 dB. The World Health Organisation classifies hearing loss into degrees of loss based on the severity of loss. The severity of loss for an ear is determined by the averages of Pure Tone Audiometry thresholds (called Pure Tone Averages) for frequencies of 500, 1000 & 2000 Hz [34]. Table 1 shows this classification.

<table>
<thead>
<tr>
<th>Degree of Loss</th>
<th>Pure Tone Average</th>
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</thead>
<tbody>
<tr>
<td>Normal Hearing</td>
<td>0-25dB</td>
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<tr>
<td>Mild Hearing Loss</td>
<td>26-40dB</td>
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<tr>
<td>Moderate Hearing Loss</td>
<td>41-55dB</td>
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<tr>
<td>Moderately severe Hearing Loss</td>
<td>56-70dB</td>
</tr>
<tr>
<td>Severe Hearing Loss</td>
<td>71-90dB</td>
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<tr>
<td>Profound Hearing Loss</td>
<td>91dB above</td>
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</table>

**Disabling Hearing Loss:** The WHO further recognizes some degrees of hearing loss as ‘disabling’. In children aged 0-14 years, hearing loss is ‘disabling’ when the Pure Tone Average in the better hearing ear is greater than 30 dB [6]. This means that some hearing loss which would normally be classified as mild may be disabling.

**Types of Hearing Loss:** The following types of hearing loss are recognized: conductive hearing loss, sensoriournural hearing loss and mixed. Conductive loss is secondary to an abnormality of the outer ear or the middle ear. The abnormality reduces the intensity of the sound reaching the cochlea. Examples of causes include occlusion of the ear canal by cerumen, middle ear infection, perforation of the tympanic membrane and ossicular chain abnormalities. Conductive loss can usually be cured medically or surgically. Sensoriournural loss results from lesions of the cochlea (sensory type) or the eight cranial nerve and its central connections (neural type). The problem here is not with conduction of sound but with the transmission or perception of neural inputs. It usually cannot be treated but adequate rehabilitation is usually possible. In mixed loss, elements of both conductive and sensoriournural loss are present in the same ear [35].

**Causes and Types of Hearing Loss in Children:** A wide range of pathologies ranging from hereditary to neoplastic can be responsible for hearing loss in children. Table 2 shows a list of causes of hearing loss in children [36]. Several studies have however shown regional variations in the predominant causes. It has been reported that in western literature about 24-39% of the causes of hearing loss are due to genetic factors [37]. In Egypt, previous studies have pointed to heredity and infection as the main causes of hearing loss [37, 38]. However, in a recent national household survey for hearing loss, the most common cause of hearing loss was otitis media with effusion which accounted for 30.7% of those with hearing impairment: the peak age group was 0-4 years followed by 5-14 years [38]. According to Zakzouk and Al-Anazy in Saudi Arabia the most common cause of hearing loss in age group 5-15 years was otitis media with effusion [39].

The following data also highlight the importance of ear infection as a leading cause of hearing loss in children. In a survey of office practice patients in the United States, the commonest sequel of acute otitis media was otitis media with effusion (OME) associated with conductive hearing loss. Around 80% of children had OME at least once before the age of four and chronic suppurrative otitis media was the cause of hearing loss in 17.6% of cases [40]. The frequency of middle ear infections account also for a high incidence of conductive hearing loss. In a study by Ossama et al. [41], 13.8% of 1600 children (0-14 years) screened were shown to have hearing loss. The majority (85.9%) had conductive hearing loss. Only 14.1% had sensoriournural hearing loss. Often children with ear infections have both ears affected. In Ossamma et al’s series 78.7% had bilateral hearing loss. The majority of patients had mild hearing loss (right-79.3%, left- 77.7%). OME was also the most common cause of hearing loss (66.5%) [41].
Table 2: Types and causes of hearing loss [36]

<table>
<thead>
<tr>
<th>Conductive deafness</th>
<th>Sensorineural deafness</th>
<th>Neural deafness</th>
<th>Psychogenic deafness</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Congenital</td>
<td>A. Congenital</td>
<td>A. Cochlear nerve injury</td>
<td>A. Malingering</td>
</tr>
<tr>
<td>i) Congenital meatal stenosis</td>
<td>i) Genetic</td>
<td>B. Cochlear nerve tumor e.g. Acoustic neuroma</td>
<td>B. Inattentiveness</td>
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<td>ii) Fusion or agenesis of Ossicular chain</td>
<td>ii) Intra-Uterine viral infection e.g. maternal rubella</td>
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<tr>
<td>iii) Otosclerosis</td>
<td>iii) Hemolytic disease of the newborn</td>
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<tr>
<td>B. Trauma</td>
<td>iv) Anoxia</td>
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<tr>
<td>i) Barotrauma</td>
<td>v) Birth injuries</td>
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<tr>
<td>ii) Foreign bodies</td>
<td>vi) Congenital syphilis</td>
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<tr>
<td>iii) Rupture of tympanic membrane</td>
<td>i) Head injuries</td>
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<tr>
<td>iv) Traumatic Ossicular Chain Disconnection</td>
<td>ii) Bomb blasts</td>
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<tr>
<td>C. Wax</td>
<td>iii) Noise exposure</td>
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<tr>
<td>D. Infection</td>
<td>C. Infection</td>
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<tr>
<td>i) Otitis externa</td>
<td>i) Measles</td>
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<tr>
<td>ii) Acute otitis media</td>
<td>ii) Mumps</td>
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<td>iii) Secretory otitis media</td>
<td>iii) Suppurative labyrinthitis</td>
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<td>iv) Chronic otitis media</td>
<td>D. Influenza Herpes Otitis</td>
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<tr>
<td>E. Tumors</td>
<td>E. Ootoxic Drugs,</td>
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<tr>
<td>i) Tumors of external ear</td>
<td>i) Aminoglycosides</td>
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<td>ii) Tumors of middle ear</td>
<td>ii) Loop Diuretics</td>
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<tr>
<td>iii) Nasopharyngeal tumor</td>
<td>iii) Antimalarals e.g. quinine</td>
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<td>iv) Cytotoxic drugs e.g. cisplatin</td>
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<tr>
<td></td>
<td>F. Others</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td>i) Meniere’s disease</td>
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<td></td>
<td>ii) Otosclerosis</td>
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**Diagnosis:** It is important to introduce intervention measures in the first few months of life. Early detection of hearing loss and treatment or introduction of intervention measures is the prerequisite first step in the process towards linguistic competence, literacy development and achievement of maximum potential [42-44]. UNHS is the method by which most developed countries achieve early detection in the newborn. Systematic screening of children is also necessary to detect hearing loss that develops later. Hearing testing in children can be done by physiologic (objective) and behavioral (subjective) testing.

**Physiologic Testing:** This makes use of objective means that do not depend on the patient’s cooperation and are not invalidated by a patient’s lack of cooperation. Screening tests utilizing Oto Acoustic Emissions (OAE) technology and Auditory Brainstem Response (ABR) audiometry are the methods used especially to detect PCEHL [45]. They are widely used in newborn and childhood hearing screening programmes [46]. Another objective means of hearing assessment is Tympanometry, a form of impedance audiometry used to test middle ear function [47].

Auditory Brainstem Responses (ABR) refer to a series of electrical potentials that can be recorded from the scalp during the first 10 to 20 milliseconds following the onset of a transient auditory stimulus [48]. ABR can be reliably obtained in infants at intensity levels suggesting normal peripheral auditory function [49-53]. They have been detected in humans neonates as early as 25 weeks’ gestational age [54] and are not affected by sleep, sedation, or attention [55]. Thus, ABR audiometry is well suited to estimation of auditory sensitivity in infants and children who, because of developmental stage or handicapping conditions, cannot be tested reliably using conventional behavioral techniques.

Otoacoustic Emissions are low-level sounds originating within the cochlea [56]. Through reverse propagation, some of this acoustic energy leaks from the cochlea and travels through the middle ear to the external auditory meatus, where it can be recorded using a sensitive microphone [57, 58]. They are detectable only when both the cochlea and middle ear systems are functioning normally or near normally.

Tympanometry measures the relative change in middle ear compliance as air pressure is varied in the external ear. Because middle ear disorders alter the shape
of the tympanogram in predictable ways, tympanometry is useful in delineating types of middle ear conductive abnormality. Given the prevalence of middle ear disease in children, tympanometry has become an indispensable tool in pediatric audiologic assessment. Tympanograms are classified as types A, B and C, according to the qualitative system [47, 59, 60]. They provide information about disorders of the tympanic membrane (perforation, scarring, retraction), tympanum (middle ear effusion or abnormal pressure) and eustachian tube function [61-63].

**Behavioral Testing:** Pure tone audiometry (PTA) is a method used in measuring hearing sensitivity across a range of frequencies. PTA evaluation assesses both degree and type of hearing loss for each ear individually. It can be used in children who are old enough to comprehend and who can hear well enough to respond to the instructions for the test. For each frequency, a pure tone signal is presented to the ear and the intensity of the signal is varied until the level at which the participant is just able to perceive the tone is identified. This level is the pure tone threshold for that frequency. A higher threshold indicates that a more intense signal is needed to perceive the tone and signifies greater hearing impairment. The amount of intensity (measured in decibels) that has to be raised above the normal level is a measure of the degree of hearing impairment at that frequency. Hearing sensitivity is plotted on an audiogram, which is a graph displaying intensity as a function of frequency [64], a plot of frequency in hertz (Hz) against intensity in decibels (dB). Even though young normal adults can hear frequencies as low as 20 Hz to as high as 20,000 Hz, the frequencies used in clinical measurement include only those from 250 Hz to 8000 Hz [35]. Pure Tone Audiometry is a behavioral test used to measure hearing sensitivity. This measure involves the peripheral and central auditory systems. Pure-tone thresholds (PTTs) indicate the softest sound audible to an individual at least 50% of the time. Figures 1, 2 and 3 show audiograms for conductive, sensorineural and mixed hearing losses, respectively.

**Conditions Mimicking Hearing Loss and Those Associated with Hearing Loss in Children:** A number of other conditions in children may mimic hearing loss. Several are child psychiatric disorders and need to be identified early and appropriately managed. These conditions include selective mutism, attention deficit hyperactivity disorder (ADHD) and even depression. Developmental disabilities like autism, specific language impairment or learning disabilities are also commonly associated with hearing loss and also need to be identified early and appropriately managed. Central Auditory dysfunction is a condition that mimics hearing loss and deserves special mention here.

**Central Auditory Dysfunction:** This is also known as central auditory processing disorder (CAPD). It is thought to affect up to 5% of school-aged children. These children cannot process sound normally because their ears and brain don't fully coordinate. There are difficulties in recognizing subtle differences between sounds in words, even when the sounds are loud and clear enough. The problems usually occur in background noise, which is a natural listening environment. Children with CAPD can usually detect pure tones that are delivered one by one in a very quiet room, for example, the sound-proof room of the standard testing environment, but have difficulty in understanding speech signal presented under less than optimal conditions. However, Audiologists can determine if a child has CAPD by interacting with the child and through auditory processing testing. If the auditory deficits are not identified and managed early, there may be speech and language delays and academic problems. However, once diagnosed, the prognosis is good with speech therapy and regular follow up visits [65].

**CONCLUSION**

A child who cannot hear will not develop speech or language and if these children do not receive attention they are sentenced to exclusion, limited achievement and poor quality lives. These children must receive attention and they must receive it as early as possible. The longer the delay, the worse the outcome. Unfortunately, a large amount of the world's children suffer silently from the quietness brought about by hearing loss. And almost all (about 90%) of them live in areas where they do not have access to facilities for early diagnosis.

This is an emergency situation that calls for urgent action. Family Physicians, primary care workers, others who deal with children, governments, governmental and non-governmental organizations should be adequately sensitized so that they may all act their parts in the primary prevention (through primary health care services), secondary prevention (through prompt diagnosis and adequate treatment of infections and screening services to facilitate early detection) and tertiary prevention (through adequate rehabilitation of affected children). The efforts that are being made at present must be sustained and strengthened through strong intersectoral collaboration.
REFERENCES


