

Human Hearing Loss

Epidemiology, Mechanisms, Interventions

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Hearing loss affects as much as 5% of the global human population and its negative consequences, often exacerbated by cultural bias or distributive injustice, include delayed cognitive and language development, learning deficits and poor academic performance, chronic unemployment and dependency, poverty, elevated risk of harm and poor health. This paper is based on a review of the academic literature as well as other credible published resources to identify the principal causes of hearing loss; its consequences for individuals, communities, and states; and potential interventions most appropriate for developing and low-resource countries where hearing loss is currently most prevalent and its burdens most egregious.

BACKGROUND

Audition, or hearing, is a type of mechanosensation in which the perception of sound is stimulated by nerve impulse transduction of pressure changes in the medium surrounding the organism. As one of the so-called five major senses, humans rely on hearing for social communication, learning, and for evaluating and orienting within the environment. Degradation in, or the absence of, this ability is referred to as hearing loss (HL) or deafness. It is estimated that approximately 360 million people, or as much as 5% of the global population, suffer from disabling hearing loss, including 32 million children ¹.

Normal human hearing is defined by thresholds of 25dB (or better) in both ears ¹. Individuals with thresholds 25dB or more above normal in one or both ears ¹ are considered to have hearing loss, which may be classified as mild, moderate, severe, or profound ¹ depending on the threshold and range of hearing. A best threshold greater than 40dB above normal (for adults, 30dB for children) characterizes disabling hearing loss ^{1,2}, which coincides with the categories of moderate, severe, and profound. Deafness properly refers to either profound hearing loss or the complete absence of hearing.

CONTEXT, BURDEN, and COST

Because human cochlear hair cells do not regenerate under normal circumstances ³, anatomical damage to key auditory anatomy can be permanent and cumulative. Adding to the tragedy, approximately half of all cases of hearing loss, regardless of cause, are preventable ^{1,2}. It thus makes sense that most hearing loss occurs in developing countries where economic resources for health care are scarce and so critical interventions at the points of prevention, diagnosis, and treatment are regularly neglected ². Further complicating the issue, non-resource, community-based factors such as social attitudes, local customs, and cultural biases have been found to lead to delays in diagnosis and again in treatment for HL individuals already limited in health care access by geographic and economic circumstances ². HL has been consistently associated with rural living conditions in developing countries, and recently with economic status, together suggesting strong interplay between socioeconomic demographics and political will in national resource allocations for diagnosis and treatment ².

The consequences of hearing loss extend beyond reduced sensation experienced by its sufferers. Those with disabling hearing loss experience impaired communication ² in all aspects of their lives. Because many important stages of cognitive development depend on hearing, the consequences may be far more severe and enduring for children. In the absence of adequate, disability-appropriate support, speech and language development – including vocabulary, sentence structure, and pronunciation skills ⁴ – may be delayed or fully occluded ². Consequently, these children are at risk for general cognitive and learning impairments, lower measured IQs, and poorer academic performance ^{2,4}. Further associations with HL include chronic illness and infections, sexually transmitted diseases, substance abuse, domestic violence, and rape ².

Harm from hearing loss may also be described in social and emotional dimensions. For example, individuals with HL often face social exclusion and / or stigmatization in the form of denied marriage rights ² or involuntary institutionalization ². Such persons may also experience limited access to services,

1 treatments, or education ¹ – whether by design, neglect, insufficient political will, or inadequate or
 2 unequally distributed resources.

3 Those with HL may be particularly vulnerable in the workplace. First, those with pre-existing HL may
 4 have reduced opportunities for employment ²; children with HL go without education, becoming adults
 5 without employable skills ^{1,2} even when none of the concomitant developmental limitations described
 6 above are present. But noise-induced, ‘on-the-job’ hearing loss may, in places where workers’ rights are
 7 poorly protected, result in termination of employment ². This, in turn, negatively affects the individual’s
 8 family and community, not just by reducing the unit’s economic input, but by potentially converting the
 9 HL-individual from economic asset to resource burden.

10 When extrapolating these themes to the national and international levels, we observe that HL is an
 11 obstacle to normal development, education, and ultimately employment, which often exposes the
 12 afflicted to extreme poverty and / or complete dependence on others ^{1,2,5}, either the family or the state.
 13 HL, therefore, impedes the larger social and economic development of communities and nations ¹ and
 14 can be seen as an important policy and development issue. Finally, HL poses a significant obstacle to the
 15 Millennium Development Goals’ target of universal primary education ⁵ and the Declaration of Human
 16 Rights’ ideal of rights to work, education, medical care, and cultural engagement ⁶.

17 **EXPOSURE LEVEL and RISK ASSESSMENT**

18 **PREVALENCE, DISTRIBUTION, and RISK**

19 As previously mentioned, approximately 5% of the world’s population, currently as many as 360 million
 20 people, have disabling hearing loss ¹, the majority of which are in low- and middle-income countries ².
 21 This includes the ~33% of those over 65 with HL, with the highest prevalence among this age group
 22 occurring in South Asia, Asia Pacific, and sub-Saharan Africa ¹. But youth is well represented among HL
 23 sufferers, too, with ~1.2 million children (ages 5-14) with either hearing impairment or total loss in sub-
 24 Saharan Africa alone ⁵. Not even the affluent escape: in the US, for example, 12-15% of school-age
 25 children have some hearing deficits attributable to noise exposure ³. The total number of HL-affected in
 26 the US is much higher ⁷.

27 Regarding risk, congenital bilateral HL in the developing world is estimated at 6 per 1,000 live births, or
 28 720,000 children with sensorineural HL born annually ², which is more than 3 times the average rate
 29 among the wealthiest nations. As many as 500 million individuals are at risk for noise-induced hearing
 30 loss ⁸, and noise accounts for approximately 37% of all adult HL cases ⁹. In the US, the risk of bilateral HL
 31 for those 12 and older is approximately 1 : 8, and 1 : 5 for unilateral HL ⁷. It’s important to note that risks
 32 vary greatly across national borders, among geographically distinct intranational populations, and even
 33 within communities with large disparities in wealth and / or healthcare access. Further, reliable HL data
 34 are simply unavailable for much, if not the majority, of the world’s population.

ETIOLOGY and MECHANISMS

There are many causes of hearing loss that vary in impact by both demographic and environmental variables. The principal causes include congenital factors (both genetic and non-genetic, accounting for approximately 38% of pediatric cases ¹⁰) and a wide variety of acquired conditions, pathogens, and exposures (accounting for approximately 30% of pediatric cases, while ~30% of cases are of miscellaneous or unknown cause ¹⁰).

CONGENITAL

In many cases, HL is present at birth or develops from conditions in the maternal or birthing environments at the rate of approximately 1-2 per 1,000 births in most developed countries. Genetics is an important component of congenital HL, as both hereditary and non-hereditary genetic factors contribute to a variety of HL-associated conditions ¹. Greater understanding of genetic contributions to nonsyndromic HL is just around the corner, as new massively parallel genetic testing methods are actively being developed ¹¹. In total, permanent childhood hearing impairment may be attributable to hereditary causes in 30-40% of cases ¹⁰. The remainder of congenital cases are primarily caused by maternal rubella ¹, which in Asia may account for as much as 40% of cases ², or complications at birth such as low birth weight, birth asphyxia, or cochlear nerve-damaging severe jaundice during the neonatal period ¹.

ACQUIRED

In the US, approximately 60% of adults over 70 experience some degree of HL, which, when compared to the 1-2 per 1,000 congenital rate, reveals that HL is primarily an acquired condition. What is happening between birth and age 70 that results in such a high prevalence of HL? Principally, infectious diseases, ototoxic and noise exposures, the natural processes of ageing, and anatomical injury or obstruction – each potentially interacting with genetic factors ¹² and exacerbated by the circumstances of poverty and insufficient access to care – are driving acquired HL.

INFECTIOUS DISEASES

Infectious diseases, including meningitis, measles, mumps, and chronic ear infections such as otitis media are the primary causes of acquired HL among children ^{1,2} in developing countries primarily because effective vaccines or interventions are widely available and routinely administered in the developed world.

Though mechanistically variable, these diseases primarily damage the cochlea organ or cochlear nerve¹³ rather than interfere with bone conduction ¹⁴, and usually early in the infection ¹³. Much of this damage can be rapidly reversed, but only with the prompt and adequate treatment ¹³ typically unavailable in resource-poor settings. These circumstances result in a double disadvantage for the poor, who, lacking vaccines, suffer a higher prevalence of infection, and lacking access to treatment, suffer unnecessarily permanent HL.

OTOTOXIC EXPOSURES

Ototoxins are chemicals that selectively damage the auditory system. Common ototoxins include prescribed drugs such as select antibiotics, loop diuretics, non-steroidal anti-inflammatories, and platinum-based chemotherapy agents^{15,16}. Some Quinines, heavy metals, and a host of environmental chemicals¹⁶ have also been shown to be ototoxic, while certain organic solvents, metals, asphyxiants, smoke¹⁷, and endocrine disruptors interact with noise exposure (discussed in the next section) to increase mechanical stress on cochlear hair cells¹⁵.

NOISE EXPOSURE

Noise exposure is a leading cause of HL across age groups¹, affecting young, adult, and elderly populations in different ways along different exposure pathways. Among working adults, HL is one of the most common occupational diseases¹⁸. Unsurprisingly, then, the workplace itself is often a primary site of noise exposure¹⁸. Music professionals, farmers, and construction and industrial workers are among those at greatest risk for work-related noise-induced HL. Outside the workplace, recreational self-exposure via electronic sound amplification is a significant problem among youth³ and adult demographics alike.

Mechanisms of noise-induced HL may be categorically divided by the type of exposure, whether acute and traumatic, or chronic. Acoustic trauma, which can result in permanent cochlear damage, may result from any acute overexposure to high intensity impulsive signals⁸. This damage may be to hair cells, to supporting structures in the organ of Corti, or to the Reissner's and tectoral membranes³. Noise induced hair cell injury is characterized by the potentially irreversible loss of cochlear nerve terminals on inner hair cells and a slow degeneration of spiral ganglion cells⁸. The extent of damage depends on the spectral and temporal aspects of the signal and the duration of exposure³.

More common is prolonged, high-intensity noise exposure. Chronic exposure can also damage hair cells of the cochlea, producing permanent hearing threshold shift and reduced speech-from-noise distinction³. But unlike acute traumatic exposure, chronic exposure may also drive metabolic changes in sensory cells³ over time, cause persistent or recurrent tinnitus, and promote the generation of reactive oxygen species in the cochlea, resulting in cumulative oxidative stress¹². Cumulative oxidative stress may be enhanced by hypoxia resulting from atherosclerosis. Further, interactions between noise and ototoxins, particularly cigarette smoke, have been associated with a higher risk of HL¹⁷.

AGEING

Age-related hearing loss, known as presbycusis, is a decline in auditory function characterized by increased hearing thresholds and poor frequency resolution¹². Though age-related HL is universal among mammalian species, the causes and mechanisms are little more than categorically resolved: cochlear ageing is a catch-all for poorly understood cellular processes among hair cells, the *stria vascularis*, afferent spiral ganglion neurons, and the central auditory pathway¹², while environmental factors (e.g., noise, ototoxins), and co-morbidities¹² (e.g., smoking, atherosclerosis) have already been mentioned. Several apoptosis gene mutations have recently been identified that may contribute to age-related HL¹⁹, and evidence of a principal role for oxidative stress-induced mtDNA damage in cochlear

cell apoptosis is accumulating¹², but the molecular complexity and dynamic interactions among contributing factors remains to be elucidated.

INTERVENTIONS

Now that we have established the harm, prevalence, and risk factors associated with hearing loss, as well as its most common causes, we may discuss what can be done to prevent, diagnose, and treat HL, with a focus on primary prevention. Long-term reduction in the incidence rate of HL begins with data collection to better define the current state of a pervasive and poorly measured condition. The dissemination of current and newly-generated knowledge and broader health systems access are also critical early steps. Targeted prevention measures in high-risk workplaces and expanded vaccinations programs will also help reduce many of the principal causes of HL among children and working adults. For unprevented and unpreventable HL, better screening and low-cost treatment options can dramatically reduce the physical, developmental, social and economic sequelae of HL.

DATA COLLECTION

There is broad consensus in the hearing loss literature, among public health experts, and reflected in the World Health Organization's topic analysis that the true global prevalence and magnitude of the economic and social consequences of hearing loss are unknown and must be established^{1,2,5,18} as a first priority. In sub-Saharan Africa, for example, almost no epidemiological data exists in the literature documenting either the prevalence or causes of HL in the region⁵.

Without a clearer global picture of the cases, causes, and consequences of HL, purposeful, systematic, coordinated, **effective and efficient** planning and action will remain stymied or impotent. There is a multiplicative property of poverty such that HL is least appreciated in the very poor regions – where birth rates and infections are high while prevention, screening, and treatment are low or non-existent – that would benefit most from strategic national and international action. There is a great need, therefore, for the international and global health communities to make rational investments in HL-focused epidemiological studies as a critical step toward amelioration.

EDUCATION and IMPLEMENTATION

Unequal distribution of knowledge is a common theme in global health. There are tremendous barriers – cultural, political, and economic – to information access for much of the world's population, and their slow, incremental removal is altering the global landscape in virtually every dimension, HL included. However, without intentional, focused efforts, this progress is unlikely to be sufficient or equitable. One information-based approach that has had great effect in other public health arenas and would likely translate well to HL prevention is the public awareness campaign, which may address HL causes, available preventive services, and / or potential treatments².

Another target for educational efforts is health care providers², who are often ignorant of or ill-informed regarding hearing assessments, language development milestones, ear exams and treatments, and patient education. Even in locations where the prevalence of HL has been measured, health care

workers have been well trained, and risk management guidelines established, there is often poor implementation of guidelines ⁵ because of low health sector buy-in, inadequate logistical or policy support, and ineffectual leadership.

PREVENTION

Hearing loss prevention efforts should take many forms to best address the disparate etiologies, myriad environments, complex risk factors, and diverse populations involved in HL. In the workplace, environmental designs that primarily reduce the generation of noise and ototoxins, and subsequently reduce worker exposure to those risks are essential. Where exposure is unavoidable, personal protective equipment – hearing protection in the form of earmuffs and earplugs – should be accompanied by proper training and compliance tracking. Unfortunately, the track record of non-pharmaceutical preventions of noise-induced HL is poor ⁸. Daily monitoring of at-ear exposure accompanied by supervisor feedback is a more promising intervention ⁸ because it requires compliance at the management level, which is rather more easily monitored and enforced than the behavior of individual workers.

Infectious diseases, which cause the majority of acquired disabling HL among children, are amenable to at least three lines of intervention. First, universal maternal immunization against rubella, routine in only 58% of countries as of 2002 ², would have a very large effect on the global prevalence of auditory and speech defects. As much as 84% of congenital rubella syndrome births result in HL and / or speech pathology ², and India's rubella and birth rates make clear the potential impact of such an intervention (India has a maternal rubella incidence rate as high as 1,000 per 100,000 births, and its annual birth rate of 22.2 per 1,000 persons means that India alone faces ~250,000 CRS-affected births per year ², of which ~80% may suffer congenital HL).

Second, immunizations against the common infectious diseases of childhood (measles, mumps, and meningitis especially) all but eliminate the risk of HL associated with these pathogens. Such vaccinations, in conjunction with proper treatment for common, unpreventable infections, especially otitis media with effusion and suppurative otitis media, could reduce HL by as much as 50% in developing countries ² – a potentially game-changing and technically achievable long-term goal.

SCREENING

Universal screening campaigns have been overwhelmingly effective in detecting hearing loss in high-income nations. But the hospital-based model (like the UNHS program in the US) may be inappropriate in countries with large numbers of home births ². While universal newborn hearing screening is a primary goal, it may only be achievable in middle- and high-income countries ⁵ because of prohibitive direct costs and the required delivery infrastructure unavailable in most developing nations. Meanwhile, at-risk hearing screening, even in high-income settings, misses between 50 and 70% of congenital HL cases, marking this approach as distinctly inferior with a poor return-on-investment (ROI) profile ²⁰.

Further, the high cost of hearing screening renders a universal screening campaign economically untenable, but dubious too. Because the costs associated with hearing screening are as much as 20x greater than the cost of full spectrum vaccination, and because rubella and other infectious agents are

leading causes of HL, investments in vaccine campaigns may provide a much greater ROI than screening in many settings ². The optimal approach, at least in some countries, will be to pair screenings with immunization programs, which has been shown to improve follow-up rates for both ² – an important point, because initial screenings are only as useful as the follow-up plan and procedures are effective ⁵.

TREATMENTS

There are two major treatment options for hearing loss: electronic hearing aids and cochlear implants, both of which face enormous logistical obstacles and are unlikely to meet demand – even if demand could be reduced to only the unpreventable congenital cases of HL. Hearing aid production currently only meets 10% of global need ². But availability is only part of the problem, as less than 50% of hearing aid recipients have been found to regularly use the devices and a full 10% don't use them at all, pointing to the need for patient education on use and maintenance of their hearing aids ².

Cochlear implants, meanwhile, remain economically out-of-reach for the majority of the world's hearing impaired, even if the technical obstacles were bridged. But even current high-cost implants have a high benefit-to-cost ratio when used among prelingual children ², despite cultural distrust of such interventional technology in some places and the deaf community's perception of threat to their identity in others ²¹. However, a low-cost cochlear implant device, if developed, could be a game changer in middle-income high-population countries like China ².

SUMMARY

Hearing loss is one of the most prevalent congenital conditions in both developed and developing countries, yet it is primarily an acquired condition resulting from exposure to infections, ototoxic chemicals, and noise. Susceptibility increases with age, and predisposing genetic factors are rapidly being identified. HL may result in delays in cognitive, speech, language, and social development, and environmental factors may extend its effects to include poor educational access and / or outcomes, low or vulnerable employment, poverty, dependency, and ultimately societal burden significant enough to impede national development efforts. Interventions should be undertaken at the level of research, education, primary prevention, screening, and treatment, with the greatest proportion of resources allocated to universal vaccination campaigns and if possible, joint screening and follow up efforts. Hearing loss – and especially its prevention – is worthy of attention from nation and international policy makers, NGOs, and the global health community because its prevalence is high, its morbidity is severe, and the potential return on investments in HL research and prevention is enormous.

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