CHILDHOOD HEARING LOSS

Strategies for prevention and care

World Health Organization

Contributors

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Contents

The interaction between a person and his or her surrounding environment is mediated through sensory experiences. The sense of hearing, in particular, fundamentally facilitates communication and fosters social interaction. Hearing is the key to learning spoken language and is important for the cognitive development of children. Without suitable interventions, hearing loss is a barrier to both education and social integration (1-10). Some 360 million people (approximately 5% of the world's population) live with disabling hearing loss[1] and nearly 32 million of them are children (12). It is estimated that over 60% of such hearing loss could be avoided through preventive measures. In addition, children who have hearing loss can benefit greatly from early identification and appropriate interventions. Action is required to ensure that the preventable causes of hearing loss are avoided and that everyone with unavoidable hearing loss can reach their full potential through rehabilitation, education and empowerment.

This document outlines the prevalence and impact of childhood hearing loss. It highlights that the majority of causes responsible for hearing loss in children can be prevented and offers strategies for action.

1. The impact of unaddressed hearing loss

While the most obvious effect of childhood hearing loss is on language development, it also has an impact on literacy, self-esteem and social skills (1-4). Untreated hearing loss is often associated with academic underachievement, which can lead to reduced employment opportunities later in life (5, 6). Communication difficulties can have lasting emotional and psychological consequences that can lead to feelings of isolation, loneliness and depression (7-10). The impact on the family is equally profound. Parents of children who are deaf or hard of hearing must deal with specific challenges, are often at greater risk of stress, have higher out-of-pocket expenses and lose more work days than other parents (13).The stress can be further exacerbated by communication difficulties with their children and increased need for support and financial resources (14-17). Untreated hearing loss also affects social and economic development in communities and countries (18).

The severity of the impact of hearing loss for a child depends on a number of factors (19-23).

[1] Disabling hearing loss in children is defined as hearing loss greater than 30 dB in the better-hearing ear.

- *Age of onset.* Children develop language in the early years of life. The impact of hearing loss on the development of spoken language is greatest in those who are born with hearing loss or develop it soon after birth (1, 24).
- *Degree of hearing loss.* Hearing loss may range from mild to profound. The greater the severity, the greater the impact (25-27). Table 1 summarizes the different degrees of hearing loss.

Table 1. Definition and characteristics of the different levels of hearing loss

Degree of hearing loss	Hearing loss in decibels (dB)	Functional characteristics
Slight/mild	26–40	The person has trouble hearing and understanding soft speech, speech from a distance or speech in a background of noise.
Moderate	Children: 31–60 Adults: 41–60	The person has difficulty hearing regular speech, even at close distances. This may affect language development, interaction with peers and self-esteem.
Severe	61–80	The person may hear only very loud speech or loud environmental sounds, such as a siren or a door slamming. Most conversational speech is not heard.
Profound	Over 81	The person may perceive loud sounds as vibrations. Speech and language may deteriorate.

- *Age of identification and intervention.* The sooner a child is identified as having hearing loss and the earlier he or she receives support, the greater the possibility of the child learning spoken language and the lower the likely adverse impact of the hearing loss (1). The Joint Committee on Infant Hearing recommends that all children with hearing loss should receive intervention by six months of age at the latest (28).

- *Environment.* The overall living environment, including access to services, significantly influences the development of a child with hearing loss. Children with access to hearing technology, special education and sign language may be able to participate at school and in social activities on an equal basis with their peers with normal hearing (in the absence of other impairments) (29).

Recent advances in newborn hearing screening, hearing technology (such as digital hearing aids and cochlear implants) and therapies that teach a child to develop spoken language through listening have greatly changed the situation for children with hearing loss. Early identification and intervention can significantly reduce the educational costs associated with hearing loss, and may improve earning capacity in later life (30-36). However, there are still millions of children who face the negative impact of untreated hearing loss in all aspects of their life (5).

> "Hard of hearing" is used to describe people with hearing loss ranging from mild to severe. At times, sounds (such as speech) are heard but not clearly understood. Such people usually communicate through spoken language and may benefit from hearing amplification with hearing aids and cochlear implants (11).
>
> Deaf children are those with severe or profound hearing loss, which implies very little or no hearing. Hearing devices, such as cochlear implants, may help them to hear and learn speech. In learning to communicate, such children may benefit from visual reinforcement, such as signs, cued speech and lipreading (11).

2. What causes hearing loss in children?

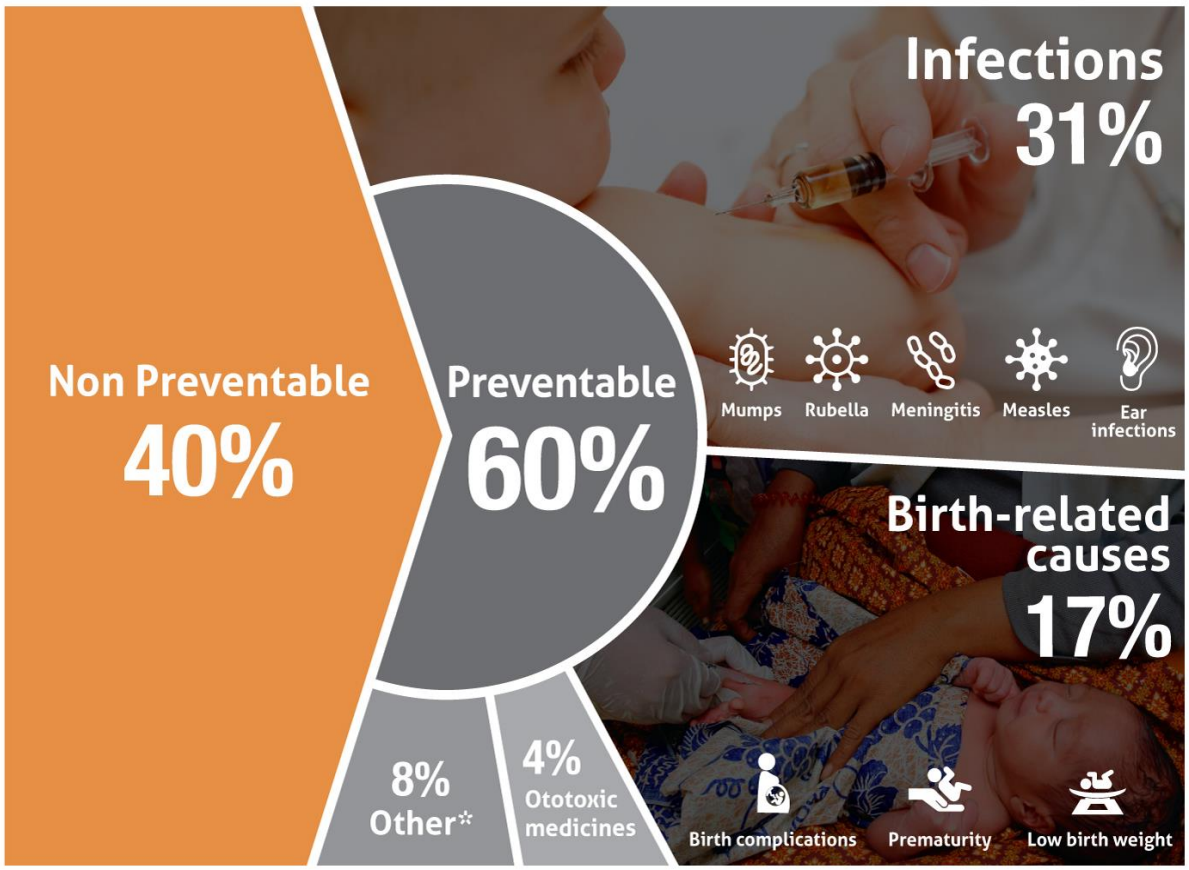
Hearing loss has many causes, and it may not always be possible to determine the exact cause. Some possible causes are listed below.

- Genetic factors. These are responsible for nearly 40% of childhood hearing loss. Hearing loss is much more frequent in children born of a consanguineous marriage (5, 37-39).
- Infections (5, 37-40).
    - Children may be born with hearing loss because the mother had an infection during pregnancy, for example, with rubella or cytomegalovirus.

- Childhood infections, such as meningitis, mumps and measles, can also cause hearing loss.
- Ear infections – often manifest as discharging or running ears, e.g. chronic suppurative otitis media (CSOM) (5, 37, 39, 43) – occur frequently in low- and middle-income countries (44). In addition to hearing loss, such ear infections can also lead to life-threatening problems, such as meningitis and brain abscesses (44).

- Conditions at the time of birth can also lead to hearing loss. Prematurity, low birthweight, lack of oxygen at the time of birth (birth asphyxia[2]), neonatal jaundice, congenital malformations of the ear and the auditory nerve (5, 39-42).
- Diseases of the ear.
  - Too much earwax (impacted cerumen), which is very common, especially in many low-resource countries (39, 45, 46);
  - Glue ear (nonsuppurative otitis media or otitis media with effusion (OME)), caused by accumulation of fluid inside the ear; this is a common childhood problem and can lead to mild to moderate hearing loss (37, 39).
- Noise (5, 37, 39, 40, 47). Exposure to loud sounds, including from personal audio systems,[3] for prolonged periods can lead to hearing loss. Even short, high intensity sounds, such as fireworks and shooting, may cause permanent hearing loss. The noisy machinery in a neonatal intensive care unit can also contribute to hearing loss.
- Medicines. Medicines, such as those used in the treatment of neonatal infections, malaria, drug-resistant tuberculosis and cancers, can lead to permanent hearing loss (ototoxic medicines). In many parts of the world, especially where their use is either unregulated or under-regulated, ototoxic antibiotics are commonly given to children to treat common infections.

[2] Birth asphyxia is the medical condition resulting from deprivation of oxygen to a newborn infant at the time of birth.
[3] A personal audio system is portable equipment, comprising headphones, earphones and a portable music player, used for listening to audio or audiovisual content. Examples include portable compact disc players and MP3 audio players.



* Other causes include: congenital non-genetic malformations and other maternal prenatal causes

Fig. 1. Overview of the causes of preventable hearing loss

3. Childhood hearing loss can be prevented

WHO estimates that about 60% of hearing loss is due to preventable causes.[4]

- Over 30% of childhood hearing loss is caused by infections, such as rubella, cytomegalovirus, mumps, meningitis, measles and chronic ear infections. Meningitis and rubella together are responsible for over 19% of childhood hearing loss. Most of these infections can be prevented by immunization and good hygiene. Ear infections and glue ear can be prevented through good ear care and general hygiene, and can be treated by prompt medical and surgical interventions.
- Complications at birth, such as lack of oxygen, low birthweight, prematurity and jaundice, account for 17% of childhood hearing loss. Such complications can be prevented through improved maternal and child health practices.

[4] This figure is based on data obtained through a meta-analysis conducted by the Prevention of Deafness and Blindness Programme at WHO. The studies used in the analysis are listed in Annex 1.

- Use of ototoxic medicines in pregnant women and children is responsible for 4% of childhood hearing loss, which could potentially be avoided.

The proportion of hearing loss due to preventable causes is much higher in middle- and lower-middle-income countries (75%) than in high-income areas (49%). The difference is probably explained by the overall higher occurrence of infections in the middle- and lower-middle-income countries and the better maternal and child health care in high-income countries.

4. Early identification helps

Early identification of hearing loss needs to be followed by timely and appropriate interventions, in order to minimize developmental delays and promote communication, education and social development (1, 23, 24, 48, 49). The choice of interventions depends on the degree and the cause of hearing loss. Otitis media can often be treated and reversed by medical or surgical means (43, 44, 50-53). Hearing loss due to other causes cannot be reversed. However, its impact can be reduced through timely use of various approaches (28, 37, 48, 54, 55):

- hearing devices, such as hearing aids, and cochlear or middle ear implants;
- hearing assistive technology, such as FM/radio systems and loop systems;
- therapy to develop spoken language, such as auditory-verbal therapy,[5] cued speech[6] and auditory-oral therapy;[7]
- development of nonverbal communication, such as sign language.[8]

Hearing screening programmes for infants and young children can identify hearing loss at a very young age (56-62). Congenital hearing loss can be detected in the first few days after birth (43, 63). Research suggests that children who are born deaf or who lose their hearing very early in life and go on to receive appropriate interventions before 6 months of age are on a par with their hearing peers in terms of language development by the time they are five years old (in the absence of other impairments) (1, 49).

[5] Auditory-verbal therapy is a method of teaching deaf children to listen and speak using their aided, residual hearing.
[6] Cued speech is a form of visual communication that combines mouth movements with cues in order to help distinguish different phonemes.
[7] Auditory-oral therapy, like auditory-verbal therapy, focuses on teaching deaf children to speak using their aided residual hearing; however there is more of a focus on speech reading and contextual cues.
[8] Sign language uses manual communication and body language to convey meaning, as opposed to sound patterns.

Children may develop hearing loss at a later age. Regular preschool and school-based hearing screening can identify hearing loss soon after its onset, allowing its adverse impact to be limited (64, 65).

Key elements in ensuring the best outcomes for children with permanent hearing loss are:

- *early identification;*
- *appropriate hearing technology;*
- *professional support for communication, learning and education; and*
- *a family-centred approach.*

For interventions to be effective, they should be appropriate, timely, family-centred[9] and undertaken through an interdisciplinary approach, which includes audiological, medical, therapeutic and pedagogical services (28, 66).

5. Strategies for prevention and care

Action is required to reduce hearing loss and improve outcomes for people with hearing loss. Governments, public health agencies, social service organizations, educational institutions and civil society groups all need to collaborate in this endeavour.

To achieve the desired results, action is needed in a number of areas.

A. Strengthening relevant programmes and organizations

- Strengthen immunization programmes (67-70), to prevent many of the infections that lead to hearing loss, such as congenital rubella, meningitis, mumps and measles. Potentially, over 19% of childhood hearing loss could be avoided through immunization against rubella and meningitis.[10]

  Action: include these vaccines in the national immunization programme and ensure widespread coverage.

[9] A family-centred approach is a service delivery model that focuses on the role of the family in the development of children
[10] Based on results from a meta-analysis (Annex 1).

- Strengthen maternal and child health programmes to prevent low birthweight, prematurity, birth asphyxia, congenital cytomegalovirus infection, and neonatal jaundice (41, 71).

  Action: improve maternal and neonatal care through

  - *improved nutrition,*
  - *awareness of hygienic practices,*
  - *promotion of safe birth,*
  - *prompt management of neonatal infections and jaundice.*

- Strengthen organizations of people with hearing loss, parents and family support groups.

  Action: encourage the formation of support groups for people with hearing loss and their families.

B. Implementation of screening and intervention programmes

- Implement newborn and infant hearing screening with tracking, and initiate appropriate interventions to identify and treat children with congenital or early-onset hearing loss (5, 24, 48, 56, 72). A newborn hearing screening programme should follow a family-centred approach, in which families are empowered to make decisions for their children (66).

  Action: put early intervention programmes in place and implement newborn hearing screening programmes (based on physiological methods) that focus on:

  - *appropriate interventions, ideally initiated before 6 months of age;*
  - *family support, including guidance and counselling of parents;*
  - *hearing rehabilitation through hearing aids and cochlear implants;*
  - *suitable therapy and communication options.*

- Implement school-based hearing screening with the aim of identifying, referring and managing common ear diseases and hearing loss (57, 59, 60, 65).

Action: integrate ear and hearing screening in school health programmes and develop links for provision of suitable medical, surgical and rehabilitative care.

C. Training

- Train primary-level physicians and health care workers about the relevance of ear diseases, the need for early intervention to address hearing loss and the available treatment options (73-75). This would allow provision of accessible services and facilitate referral for management of ear diseases and hearing loss. WHO's training manuals on primary ear and hearing care (76) and its manual on promoting ear and hearing care through community-based rehabilitation (77) are useful resources for this.

  Action: establish training programmes in primary ear and hearing care for primary-level health care providers.

- Train otologists, audiology professionals, other medical professionals (such as nurses), therapists and teachers to provide the required care and services. This is an important step in addressing ear and hearing problems.

  Action: set up professional training programmes to develop the appropriate workforce in the field of hearing health and education for persons with hearing loss.

D. Making appropriate technologies accessible

- Make hearing devices accessible (44, 78-80). Advances in the field of hearing aids and cochlear implants have considerably improved the available options for people with hearing loss. Despite this, only a fraction of those who need these devices have access to them (81). Particularly in developing countries, there are several significant barriers to access to hearing aids for people with hearing loss. A major barrier is the cost of hearing aids, batteries and maintenance (78, 80, 82). There is also a scarcity of health care professionals able to fit, maintain and repair devices (78, 80, 82). Transportation costs and travel time to a health centre may be prohibitive for many people with hearing loss, especially in rural areas (78, 82). Technological advances, such as solar-powered or self-fitting hearing aids, may help to overcome some of these significant barriers in the future (78, 83).

Action: develop sustainable initiatives for affordable fitting and maintenance of hearing devices, and provide ongoing support for people using these devices.

- Make communication and education accessible (84, 85). A deaf child benefits greatly from early introduction to language. This may be in the form of rehabilitation for verbal communication (such as auditory-verbal and auditory-oral therapy). Policy-makers should also promote alternative means of communication, including sign language, total communication,[11] bilingual/bicultural (bi-bi) teaching[12], cued speech[13] and lipreading approaches (86). Use of loop and FM systems in classrooms and public places, as well as provision of captions on audiovisual media, are important for improving accessibility of communication for people with hearing loss.
  Action: ensure access to communication through all available means, in consultation with all stakeholders, including people with hearing loss and their family.

E. Regulation and monitoring

- Regulate and monitor the use of ototoxic medicines, in order to minimize the dangers posed by their indiscriminate use (87, 88). Where their use is unavoidable, regular audiological monitoring will help identify hearing loss at an early stage.
  Action: develop and implement legislation to restrict the sale and use of ototoxic medicines; sensitize health care providers to the need to conserve hearing in people taking these medicines.

- Regulate and monitor noise levels in the community, especially at recreational venues and sports arenas. The addition of safety features to personal audio systems can help to reduce the risk of hearing loss associated with their use (89).
  Action: develop and implement regulations regarding environmental noise, including at recreational venues; implement standards for safe listening devices.

[11] Total communication incorporates all means of communication: formal signs, natural gestures, fingerspelling, body language, listening, lipreading and speech.
[12] A philosophy of teaching that recognizes the authenticity and importance of both hearing and deaf cultures and that incorporates elements of both in the classroom.
[13] Cued speech is a visual communication system — mouth movements of speech combined with "cues" to make all the sounds (phonemes) of spoken language look different.

F. Raising awareness.

- Raise awareness about healthy ear care practices that can reduce ear infections (44, 90). For instance, avoiding insertion or instillation of any substance (e.g. a cotton bud) into the ear can help to decrease ear problems. Ensuring that children with ear pain are not treated with home remedies and consult a medical practitioner can prevent chronic ear infections and the associated hearing loss.
  Action: establish awareness programmes promoting ear and hearing care in the community.

- Raise awareness about the dangers of loud sounds by educating children at an early age about the risks associated with high volumes, especially in a recreational context (firecrackers, loud music, use of headphones, noisy games) (47, 89, 91). This can help to modify behaviour patterns and promote safe listening, which in turn can prevent the development of noise-induced hearing loss during childhood and adolescence or later in life.
  Action: develop and implement awareness programmes targeting young children with the aim of promoting safe listening habits.

- Raise awareness among the community as a whole in order to reduce the stigma associated with hearing loss. Highlighting and sharing success stories about people with hearing loss can be effective in reducing stigma associated with hearing loss, hearing devices and alternative communication methods (77).
  Action: engage role models to raise awareness about hearing loss prevention and care.

Strategic planning around the above actions can help reduce hearing loss and its adverse impact on those who live with it. In line with the principles of the United Nations Convention on the Rights of Persons with Disabilities, improved hearing and access to communication facilitates education and employment, and fosters social inclusion and psychological well-being among people with hearing loss (92). Many countries have already started implementing such strategies and have established models for prevention, identification and intervention.

Today, the causes of hearing loss are known and preventive strategies have been identified; technology is available to detect hearing loss at the earliest stage of development, and intervention techniques are well established. Thousands of children with hearing loss are acquiring communication skills, and will have the same opportunities in life as their hearing peers. On the other hand, millions are still facing the adverse consequences of hearing loss throughout their life.

Case studies

Cambodia

Mom Srey Piseth is an eight-year-old girl living in rural Cambodia (93). Like many other children in Cambodia, Piseth suffers with ear infections, which cause discharge from both her ears. Ear discharge is very common among children in rural Cambodia, and is often considered normal. However, it causes hearing loss, which may lead to devastating long-term effects on communication, language development, and educational progress. If left untreated, it can lead to serious medical complications and may even be fatal.

Piseth's problems were discovered by an outreach medical team. Her condition was so advanced that not only had she lost most of her hearing, the disease had also started to destroy the bone in her skull. She underwent immediate surgery to remove the infected tissue and bone. Following the surgery, Piseth's ear improved and she went back to her village and school. Her progress is being carefully monitored by the medical team.

Thailand

Congenital rubella syndrome (CRS) can cause hearing loss, eye and heart defects, and other lifelong disabilities, including autism, diabetes and thyroid dysfunction. CRS has had a big impact on the lives of a Thai family from Bangkok (94). When Chi was pregnant with her daughter Im, her husband became ill, and had a skin rash – the classic presentation of rubella. Chi also fell ill with the same symptoms a few days later. She went to the doctor and was told she would be fine. However, at the time she was unaware that she was one month pregnant.

After Im was born, her parents realized that she had problems with her vision. It soon became clear that she also could not hear. "Im is deaf," Chi explains "she can neither hear nor speak." CRS, resulting from the rubella infection during pregnancy, is the most likely cause of Im's problems. Chi hopes that, with good rehabilitation, her daughter will be able to lead a healthy and happy life.

The highest risk of CRS is in countries where women of childbearing age do not have immunity to rubella, either from vaccination or from having had the infection. Large-scale rubella vaccination during the past decade has practically eliminated rubella and CRS in

many developed and in some developing countries. In April 2015, the WHO Region of the Americas became the first region to be declared free of endemic transmission of rubella (11).

United Kingdom

Lindsey contracted cytomegalovirus (CMV) infection when she was pregnant with her daughter Charlie (95). Soon after birth, Charlie failed her hearing test and it was confirmed that she was deaf in her left ear. When Charlie reached three years of age, the hearing in her right ear also deteriorated. The CMV infection contracted by Lindsey during her pregnancy was determined as the cause. Thanks to appropriate interventions, Charlie has now grown into a bright young girl who attends primary school and manages very well, proudly wearing a glittery pink hearing aid.

CMV infection is an important, but relatively unknown, cause of hearing loss. The Centers for Disease Control and Prevention (CDC) in the USA estimates that about one in 150 children is born with CMV infection and that about one in five of those infected will develop permanent problems, such as hearing loss or developmental disabilities. CMV is spread by close contact with the body fluids (saliva, urine) of an infected person. Newborn CMV infection can be completely avoided through counselling pregnant women regarding the sources of infection and hygienic practices, such as regular handwashing, avoiding sharing food, avoiding contact with saliva, for example while kissing a child, and cleaning surfaces that come into contact with urine or saliva.

Uganda

Sign language has had an important positive impact on the life of Patrick Otema, a young man from a remote area of Uganda. Patrick was born deaf in an area where there were no schools for deaf children, and spent most of his childhood without knowledge of sign language and thus without any communication. Patrick spent most of his days alone in his hut, isolated from the world.

The Uganda National Association for the Deaf, a nonprofit organization dedicated to empowering individuals with hearing loss, arranged for Patrick to take his first sign language class when he was 15 years old. These classes transformed Patrick's life. He is still taking classes today and hopes to teach other deaf people in the future. A documentary called "15

and learning to speak" was made about Patrick's experience and broadcast on the BBC Channel 4 programme, *Unreported world* (96).

Viet Nam

Ngoc was born in Viet Nam; soon after she was born, her family noticed that she was not responding to sounds around her (97). When Ngoc was 15 months old, her parents took her to the doctor for a hearing test, where it was confirmed that she had severe hearing loss. Ngoc's family was devastated as they had no idea how to deal with hearing loss. The doctor recommended hearing aids for Ngoc and referred the family to an educational programme for children who are deaf or hard of hearing, to get additional information.

Ngoc was fitted with a pair of hearing aids when she was 17 months old. She was then enrolled into an early intervention programme, where she is making great progress, learning to listen and speak. The combination of appropriate hearing technology, locally based care in audiology, auditory-verbal therapy from locally based trained professionals, and family support have contributed to her success.

Canada

One night while Marco was sleeping in his mother's arms, her husband picked up a brass bell and shook it continuously; Marco did not budge. That is when they knew that something was not right (97). The next week, a specialist at the children's hospital diagnosed Marco's severe-to-profound bilateral hearing loss. Marco was enrolled in a programme where he would learn to listen and speak. He received his first pair of hearing aids and started walking, all by the time he was 10 months old.

The curious little boy loved to listen and spent hours with his big sister, colouring and talking. Marco was integrated into mainstream school and graduated as an honour student. He is now in his third year of a mechanical engineering programme. Marco is an inspiration to all who have met him and he is proud to say that he continues to overcome any challenge set before him.

United States of America

Josephine failed her initial auditory brainstem response screening at birth in the USA and was diagnosed with a bilateral severe-to-profound hearing loss (97). She was fitted with hearing aids immediately. However, as the hearing aids did not give her any benefit,

Josephine received a cochlear implant when she was one year old. She received therapy from the age of eight months, and her latest evaluation revealed normal language skills and mildly delayed speech skills compared with normal hearing children. She now attends a mainstream preschool and continues to receive individual speech therapy lessons to improve her articulation skills. Soon, Josephine will join other neighbourhood children at the local primary school.

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Annex 1. Articles used in the meta-analysis on causes of childhood hearing loss.

A meta-analysis was performed to estimate the amount of childhood hearing loss that is considered preventable. We intend to publish this meta-analysis separately.

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