

OPEN ACCESS GUIDE TO AUDIOLOGY AND HEARING AIDS FOR OTOLARYNGOLOGISTS



AUDIOLOGY FOR SPECIAL POPULATIONS: INFANTS

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The *World Health Organization (WHO)* defines disabling hearing loss in children as $>30\text{dB HL}$ in the better hearing ear, based on pure tone audiometry and a pure tone average (PTA) at frequencies 500Hz, 1000Hz, 2000Hz and 4000Hz. The *WHO* reported in 2013 that approximately 360 million people have disabling hearing loss of which 32 million are children¹. The majority lives in low and middle income countries. Half of all cases of hearing loss are avoidable through primary prevention *e.g.* immunisation programs and promotion of safer childbirth². Children with hearing loss may benefit from a range of interventions such as hearing aids, cochlear implants, captioning of text, sign language training and educational and social support.

Prevalence of hearing loss in children

The prevalence of hearing loss in developed countries is 1-2/1000 births. In Africa the prevalence is 6/1000 births *i.e.* 500 new cases per day (455 if corrected for infant mortality). According to the *WHO* 15.3 million African children <15 years of age have a hearing loss of $\geq 31\text{dB}$ ^{1,3}.

Large numbers of children with hearing loss who need help and counselling are in developing countries where there is an alarming paucity of ENT specialists, audiologists and speech therapy services⁴. In the developed world the ratio of audiologists per general population is typically 1:20 000 as compared to Africa where it typically 1:1 million. A contributing factor is that there are inadequate training centers for audiologists^{4, 5}. The responsibility therefore generally rests with the local ENT doctor or clinical medical officer to diagnose hearing loss, to counsel parents and teachers, and to provide rehabilitation.

Impact of hearing loss

Functional impact

One of the main consequences of hearing loss is the ability to communicate with others. The development of spoken language is often delayed in children with hearing loss or absent in total deafness. Hearing loss and ear diseases such as otitis media can significantly affect academic performance. However, when opportunities are provided for people with hearing loss to communicate through spoken/written/sign language then they can participate on an almost equal basis with others.

Social and emotional impact

Exclusion from communication can severely impact everyday life, causing feelings of loneliness and isolation. The feeling of not understanding and not being understood is extremely frustrating for a child.

Economic impact

In developing countries, children with hearing loss and deafness rarely receive any schooling. Adults with hearing loss have a much higher unemployment rate in developing as compared with developed countries. Improving access to education and vocational rehabilitation services, and raising awareness of hearing handicap especially among employers, would probably decrease unnecessarily high unemployment rates among hearing impaired adults¹.

Risk factors associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood

Congenital

Congenital hearing loss is present at birth or may be acquired thereafter. It can be caused by genetic syndromes or nonsyndromic factors, or environmental factors such as complications during pregnancy and childbirth. Many congenital causes are unknown (~30-50%). Common factors include:

- Family history of permanent childhood hearing loss
- Infections during pregnancy *e.g.* rubella, syphilis and cytomegalovirus
- Low birth weight (<1500g)
- Birth asphyxia (lack of oxygen)
- Maternal alcohol and drug abuse during pregnancy
- Ototoxic drugs taken during pregnancy *e.g.* aminoglycosides and antimalarials
- Severe neonatal jaundice; this is a major risk factor in sub-Saharan Africa
- Craniofacial anomalies including of the pinna and ear canal. This may be evident from ear tags and pits and radiological temporal bone anomalies (*Figure 1*)
- Physical findings associated with syndromes *e.g.* Down's, Pendred's, Turner's, Alport's, Usher's type 1, CHARGE, Waardenburg's (*Figure 2*), and Jervell Lange-Nielsen syndrome

Acquired

Acquired hearing loss can occur at any age

- Wax or foreign bodies blocking the ear canal can cause conductive hearing loss
- Collections of fluid in the middle ear (otitis media with effusion/OME) can cause transient conductive hearing loss (*Figure 3a*)



Figure 1: Craniofacial anomaly with atresia of external ear canal



Figure 2: Waardenburg's syndrome

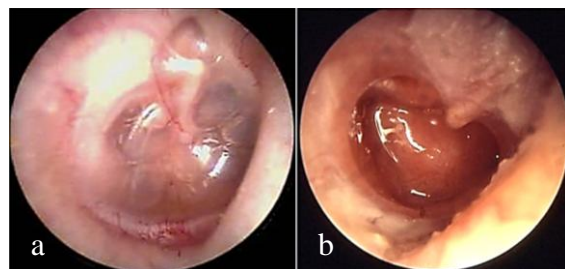


Figure 3 (a): OME with retracted tympanic membrane and serous fluid in middle ear; (b): Discharging ear with central tympanic membrane perforation

- Chronic ear infection commonly presenting with otorrhoea or leading to serious, life-threatening complications such as meningitis, lateral sinus thrombosis or brain abscesses (*Figure 3b*)
- Infectious diseases *e.g.* mumps, measles, and meningitis can cause hearing loss in childhood, but also later in life
- Ototoxic drugs *e.g.* antibiotics, immunosuppressives and antimalarials can damage the inner ear (hearing and/or balance)⁶
<http://emedicine.medscape.com/article/857679-overview>
- Head injury, especially injury to the skull base and middle and inner ear, can cause mild-to-severe hearing loss

In developing countries, chronic suppurative otitis media (CSOM) is the leading cause of hearing loss in children. It is mostly due to central tympanic membrane perforations and causes a conductive or mixed hearing loss. CSOM with cholesteatoma also occurs in children.

Bacterial meningitis is a serious infectious disease of childhood and is a common cause of postnatally acquired hearing impairment. Meningitis-induced hearing impairment is often rapid in onset, bilateral and severe. It can lead to sclerotic obliteration of the cochlea which may prevent a child from subsequently having a cochlear implant fitted. Cochlear implant surgery in such cases should therefore be done as soon as possible.

Streptomycin (tuberculosis), quinine and chloroquine phosphate (malaria) may destroy neural elements of the inner ear. Quinine is given intravenously in acute attacks of malaria; it is sometimes difficult to monitor drug levels and both tinnitus and bilateral symmetric and severe hearing loss may ensue⁶. There is discussion whether hearing loss is due to malaria *per se* or the treatment thereof.

Prevention

Many cases of hearing loss can be prevented through primary prevention. Simple strategies for prevention include:

- Immunizing children against childhood diseases *e.g.* measles, rubella, mumps and meningitis (haemophilus influenzae type B)
- Immunizing adolescent girls and women in the reproductive age against rubella
- Screening for and treating syphilis in pregnant women
- Improving antenatal and perinatal care, including promotion of safe childbirth
- Avoiding ototoxic drugs unless prescribed and monitored by a physician
- Avoiding ototoxic eardrops
- Referring babies at high risk for hearing loss for early assessment of hearing, prompt diagnosis and appropriate management *e.g.* family history of deafness, low birth weight, birth asphyxia, jaundice or meningitis
- Raising awareness of symptoms and signs of ear disease *e.g.* otalgia and otorrhoea, and education about ear hygiene; because early detection followed by appropriate medical or surgical interventions are keystones in prevention of hearing loss in otitis media, children with acute otitis media and chronic ear disease should be seen at health clinics

Identifying hearing loss

Early detection and intervention is the most important means to minimize the impact of hearing loss on a child's development and education. The poorer a child hears, the more it affects language and speech development and the ability to communicate.

Preschool screening for ear disease and hearing loss can be effective for early identification and management of hearing loss. If a child has a severe or profound hearing loss, parents will usually suspect it early; but if it is mild-to-moderate it may take time before it is discovered and diagnosed. The average age for detection of hearing loss/deafness is just over 2 years if a neonatal hearing screening program is not in place. A child may be 4-5 years old before hearing impairment is detected.

Managing hearing loss

Children with hearing loss may benefit from aural rehabilitation, speech therapy, hearing aids, cochlear implants, additional signing and special schooling.

Ensuring access to affordable, properly fitted, hearing aids and follow-up services in will benefit many children with hearing loss¹. However the lack of services to fit and maintain hearing aids, and access to batteries are barriers in low-income settings.

Children with deafness should be given the opportunity to learn sign language together with their families. In developed countries deaf children are offered cochlear implants, but in lower income countries there are fewer resources and it is more difficult to raise money needed for implantation (See Chapter: [Cochlear Implants in Developing Countries](#))

How to test hearing in infants

It is of course of great importance to have basic audiological equipment and to be trained to assess a child's hearing. Although one cannot always obtain reliable hearing thresholds, one can do a lot with simple tools to get an idea of what a child

can actually hear. Combining ≥ 2 tests improves the accuracy.

History

First of all, listen to the parents' story about the child. Ask relevant questions depending on the age of the child *e.g.*

- Why do the parents suspect hearing loss?
- Can the child hear from a distance or only when the parents talk close to the ear?
- Does the child depend on sight more than hearing for communication?
- What are the language and speech milestones like? (*Table 1*)
- Is there a history of ear infection, meningitis or antibiotic treatment?
- Has the child been treated for malaria?
- Is there a genetic story of hearing loss in the family?
- Are the parents related?
- Was the pregnancy uneventful?
- Has the child been exposed to any risk factors?

Hearing, speech and language milestones

To assess a child's hearing it is important to know the normal pattern of language and speech development. *Table 1* summarises milestones for a normal hearing child; it is simplified version based on the scales of development of the *American Speech Language Hearing Association (ASHA)* and "*Listen Learn and Talk*" edited by *Cochlear*⁷. Delayed speech and language development should alert one to the possibility of hearing impairment⁸. Speech and language development differs greatly from child-to-child and depends not only on hearing, but also on other factors. It may be influenced by heredity, temperament of the child and the amount of linguistic stimuli the child has received. It may also be affected by general developmental

delays. Delays in speech and language development may also be the first sign of other pathology such as autism and attention deficit/hyperactivity disorders (ADHD).

<http://www.asha.org/public/speech/development/chart.htm>

Age	Milestones
3 months	Babbles and reacts and listens when parents talk to child
4-6 months	Moves eyes in direction of sounds. Note that deaf children also babble but that a child with normal hearing has a more speech-like babble
7-10 months	Turns and looks in direction of sound. Can produce canonical syllables such as “da”, “na”, “be” and “yaya”
10-15 months	Starts paying attention to sounds from a distance. Imitates simple sounds and words. Starts to identify persons, a few parts of the body and toys. Starts to repeat words. Likes to move rhythmically to music
16-18 months	Imitates words and understands simple questions. Points to body parts when you ask them to do so
18-24 months	Can say 2-word combinations and knows approximately 50 words and learns new words every day
2 - 3 years	Imitates words and understands simple questions. Points to body parts when asked to do so

Table 1: Hearing, speech & language development in infants with normal hearing

Distraction test

A child’s response to sound and turning and localizing a sound source can be utilised to assess hearing between ages 6-12 months. Thereafter children usually begin to inhibit these responses although the test may be used beyond this age but with some difficulty.

Technique

- Two people, a distractor and assistant, are required for the test in addition to the parent and child
- Position the child on the parent or caretaker’s lap, supported at the waist

- The assistant is positioned out of sight behind the child
- Position yourself (distractor) in front of the child
- Establish eye contact with the child
- Distract the child with a simple toy or something similar so that you get the attention of the child; an otoscope light can be used to distract a handicapped or visually impaired child or to focus a child’s attention
- The assistant presents a sound 1 meter behind and at the level of one of the ears
- The distractor observes and judges changes in the child’s state of activity
- If one is unable to find an assistant, then one can conduct the test alone by having the parent/caretaker distract the child with a toy. Stand behind the child and present different sounds. Even though one cannot observe the reaction in the child’s eyes, note if it turns towards the sound
- Use different precalibrated sound sources with high, middle and low frequencies (*Figure 4, Table 2*)
- To increase sound intensity, move closer to the child *e.g.* a spoon in a cup has a sound level of approx. 80dB at 1m, 86dB at 0,5m and 92dB at 0,25m. Talking voice *ad concham* is 60-80dB
- Don’t come closer than 20cm, as the child may then detect movement instead of responding to sound



Figure 4: Examples of sound sources

Sound source (Figure 4)	Frequency	dB
High frequency rattle	>4kHz	50-60
Wooden rattle, castanets, maraccas	4kHz	80-85
Spoon in a cup	4kHz	80
Drum with wooden stick	65Hz-4kHz	85-90
Drum with soft stick	65Hz-1kHz	85-90
Drum, soft		65dB
Rustling plastic bag	All	60-65
Cotton pulp balls in plastic box	Low	60
Talking voice		40-60
Whispering voice, soft		15-30

Table 2: Approximate sound levels with different sound sources at a distance of 1m

Whispering & conversation tests (Table 3)

At 8-9 months one can use whispering sounds like “pst”, “psch” and “yaya” delivered 3m behind the child. If one gets a reaction then hearing is normal; if there is no reaction, repeat the same sounds more loudly. If still no reaction, move closer-and-closer until the child turns or listens. You can also raise the volume of your voice. If the child does not hear a loud “yaya” sound at a distance of 0,5m, the hearing loss is at least severe.

When the child is >12 months old one can ask questions like “where is mother, father, the lamp, the nose, eyes, ears” etc. The child may point or even repeat the words. Talk to the child from behind. Do you get responses? Is it important for the child to establish eye contact to understand? Does the child like music? Sing or let the parent or caretaker sing and see if the child moves to the rhythm of the music.

The older the child, the more you depend on speech and language development. A poster with different pictures or memory cards can be helpful. Let the child tell you the name of different pictures and then ask for the pictures. Practice a few words close to child. Stand 2-3m from the child and

keep your hand or a sheet of paper in front of your mouth so that the child cannot lip-read. Whisper a question e.g. “where is the dog”? If no answer, move closer or raise your voice. How close to the ear do you need to talk to get a response? If you don’t get any answers the child may be hearing impaired. Using this method you can get a rough idea of the child’s hearing.

Note: If a child does not cooperate it could point to something else in the child’s development that is abnormal.

You can try this method in children with normal hearing and make your own references in this way.

Hearing	M4 in best ear	Description	Action
Normal	≤25dBHL	No/very minor problems; can hear whispering	None
Mild	26-40dBHL	Hears and repeat words spoken with normal voice at 1m	Counseling, possibly hearing aid
Moderate	41-60dBHL	Hears and repeats words spoken with normal voice at 1m	Hearing aid
Severe	61-80dBHL	Hears some words when shouted just beside ear	Hearing aid, lip reading, sign language
Profound	>80dBHL	Cannot hear/ understand even when shouting beside the ear	Strong hearing aid, lip reading, sign language or cochlear implant

Table 3: WHO proposed a classification of hearing loss based on M4 (Thresholds at 4 frequencies / PTA i.e. 0.5, 1, 2, and 4 kHz)

Prerequisites for audiometry in infants

Figures 5a & b illustrate the typical layout of a testing room. Verify that the test room is quiet and that there are no disturbing sounds from outside the room. The child sits at a table in front of the tester. The child must not see the audiometer/keypad; it should be hidden by a screen. The loudspeakers are placed on either side of the child at a distance of approx. 50 cm from the child’s head⁹ and must be calibrated for that distance with a sound

level meter. The audiometer, loudspeakers and headphones should be calibrated once a year to ascertain that the levels are correct. Check that the sound is emitted from the loudspeakers and/or the headset. TV screens or toy-boxes are placed on both sides to show pictures in the *Visual Reinforcement Audiometry (VRA) tests*.



Figure 5: Child's view (a); Tester's view (b)

It is not always easy to test a child's hearing. One needs at least some audiometry experience. The sensitivity of a child's reactions depends on how alert and focussed the child is and how it pays attention. It is very important to be calm and gain the confidence of the child. Never test a crying, upset or angry child. It is also important that the sound is interesting and appealing to the child. One cannot keep on testing for a long time as a child starts

getting tired. Sometimes it is better to stop and to repeat the test at another time.

Behavioural observation audiometry

From 6-12 months one can use observation audiometry. Children usually stop reacting or responding to this technique after 12 months, although the test may be tried above this age, but with some difficulty. This test requires an audiometer with loudspeakers connected to it (Figure 5). The tester observes the child's responses while presenting a series of frequency-specific auditory stimuli. The child responds to sound by turning and locating the sound source.

Technique

- Place the child on parent/caretaker's lap
- Position yourself in front of the child so that you make eye contact with each other
- Place a simple toy or an interesting object on the table in front of the child to attract his/her attention
- Present a warble tone at 1000Hz at 30dB; the duration of the tone should be about 2 seconds. A warble tone is a frequency modulated tone. If it is not possible to use a warble tone with your audiometer, you can use a pulsating tone
- Observe and evaluate the changes in the child's state of activity
- One clear response is sufficient as children get tired very quickly
- If the child does not react, increase the sound level to a maximum of 90dB or to the upper limit of the audiometer until you see some response
- Repeat the test at 2000Hz, 4000Hz and 500 Hz

Note that this method does not measure true thresholds. Younger children require higher sound levels to react than older children. The true threshold can actually be 10-15dB better than the tested intensity. Thresholds of 30dB are normal for 6-12 month's old infants so one does not need to go lower than 30dB.

Visual reinforcement audiometry (VRA)

Between 10 months and 2½ years one can use VRA. A picture is presented on a screen or in a toy-box on the same side as from which a sound is presented. The child is supposed to look at the picture or a moving toy in a box after they have heard a sound (Figure 6).



Figure 6: VRA with toy-box

Technique

- Place the child on the parent's lap
- Position yourself in front of the child so that you make eye contact with each other
- Condition the child by presenting readily audible warbler tones ~60dB at 1000Hz from one of the loud speakers while simultaneously presenting a picture or activating the toy-box on the same side as the sound
- Once the child understands the link between the image and the sound you

can start determining hearing thresholds

- Start at 25dB if you suspect minimal or mild hearing loss. If you think there is moderate-to-severe hearing loss, start at 40dB or even higher if you think there is a severe-to-profound hearing loss
- Increase the sound level until the child reacts, and then present an image as a reward
- Repeat the test at 2000Hz, 4000Hz and 500 Hz
- Do the same for the other ear
- If the child is comfortable and willing to continue you can also test at 3000Hz, 8000Hz and 250Hz
- Depending on the type of audiometer and the experience of the tester, one can switch ears to see if the child turns to the speaker from where the tone is presented
- Do not take too long time with the test as the child may get bored and want to do something else
- One may need to give the child a clearly audible tone to ensure it is alert and concentrating

Note that even at this age one does not measure true thresholds, because the child needs to hear, care about the sound, and then show a reaction. The true hearing threshold could be 5-10dB better than the "reaction-threshold" so there is no need to go lower than 25dB. Background noise in an ordinary quiet room is often > 30dB. To test lower sound levels one needs to use a sound-treated or very quiet room.

Play audiometry with loudspeakers

Children between 2-2½ years are able to participate in play audiometry but are not always comfortable with headphones. If they are comfortable with headphones one can do the test with loudspeakers instead.

Technique

- Place the child on the parent's lap
- Position yourself in front of the child so that you make eye contact with each other
- Condition the child to move a toy block or something similar every time he/she hears a sound from the speaker
- Instruct and train the child with a clearly audible warbler tone emanating from the speakers at 1000Hz or e.g. from a doorbell (*Figures 7a,b & 8*); every time they hear a sound they should be shown to move the object
- Ensure that the child understands what to do it and give a lot of confirmation and encouragement
- Start testing with a warbler tone directed at one ear at a low intensity
- Increase the intensity until the child moves the block or demonstrates in another way that the sound was heard
- Record the response on the audiogram sheet
- Repeat the tests at 1000Hz, 2000Hz, 4000Hz and 500 Hz
- If the test takes too long the child can become uncertain or bored; one must then present a clearly audible tone to alert the child
- Repeat the test for the other ear
- If the child is comfortable and willing to continue you can also test at 3000Hz, 8000Hz and 250Hz.

Unilateral hearing loss cannot be diagnosed until one can test the child with play audiometry and with headphones. A limitation of testing with loudspeakers is that one measures overall (free field) hearing; even if one ear is deaf one can elicit reactions due to cross-hearing. One may also elicit a reaction when switching between speakers. One can add to the test by shaking a rattle behind one of the ears and see if the child turns towards the

sound. This may provide a clue whether it is a bilateral or unilateral hearing loss.



Figures 7a, b: A boy is trained for play audiometry in free field (FF); he is waiting for a sound and is praised when he moves the block



Figure 8: Conditioning a child with a doorbell

Play audiometry with headphones

This method can be used from the age of 3-4 years and when the child is comfortable with headphones (*Figure 9*).



Figure 9: Play audiometry with headphone

Technique

- Place the child on the parent's lap
- Position yourself in front of the child so that you make eye contact with each other
- Train the child with loudspeakers before using headphones
- Condition the child to move a toy-block or something similar every time he/she hears a sound from the loudspeaker
- Put the headphones on and pull the cheek slightly forward to avoid the earphones compressing and occluding the ear canals
- Start testing the best ear
- Present clearly audible pure tones at 1000Hz at 40dB for suspected normal hearing; otherwise start at ≥ 60 dB
- Once the child understands, test the hearing at 1000Hz, 2000Hz, 4000Hz, 8000Hz and 500 Hz
- Determine the hearing thresholds by increasing the loudness in 5dB steps until the child moves the block

- Decrease with 10dB increments and increase with 5dB increments again to ensure that one again obtains the same threshold
- If the child is uncomfortable with the situation and has difficulty cooperating one must provide a lot of encouragement and look into the eyes of the child for some kind of reaction
- Constantly provide confirmation and encouragement until the child moves the block
- One has to conclude with one clear answer as one does not have that much time before the child gets tired and stops listening
- If the child is comfortable and willing to continue one can also test at 250Hz and 3000Hz. There is no need to test at intensities below 20dB

Tympanometry

Tympanometry is a useful complementary test to confirm the status of the middle ear. (See Chapter: [Tympanometry](#))

Otoacoustic emissions - OAE

OAE is an objective way to assess a newborn or young child's hearing. If one obtains emissions you know that the hearing (outer hair cell function) is normal or near-normal (< 30 dB hearing loss). If you are unable to measure any emissions there could be a hearing loss. It is very important that the child is calm and quiet during the test and that the state of the middle ear is normal e.g. no middle ear effusions. This should be confirmed with otoscopy and/or tympanometry before commencing OAE tests. See Chapter: *Objective Hearing Assessment: Otoacoustic emissions*

How to intervene and counsel

- Identify and test the hearing of children with risk factors as early as possible
- The earlier one fits a hearing aid the better for hearing and speech development and for oral communication
- Refer the child to a hearing center that can select and fit a hearing aid
- Hearing impaired and deaf children need all the external input they can get
- Counsel the parents that they need to talk a lot to the child from a close distance. They need to explain a lot to the child and use colourful language. They should use proper words for things, not “it”. The child needs to hear words over and over again before it can start using words him/herself
- It is always preferable for the child to be at a close distance to its mother or caretaker because it can then pick up sounds easier
- It is crucial for the child to be a part of its community and to participate in everyday life activities
- Explain that a child with hearing loss often gets frustrated when they do not understand or are not understood
- The child needs to properly see the face and lips of the person who is speaking. Ensure that there is enough light on the face of the speaker
- When severe or profound hearing loss is detected late, start by letting the child “feel” the sound you are producing *e.g.* blowing on the hand when pronouncing “ppp”, vibrating on the neck when pronouncing “mmm” or on the nose when pronouncing “nnn” (*Figure 10*)
- Encourage the child to play with a bird feather, a small cotton bulb or a balloon and let the child blow it forwards by blowing with a “p” (*Figure 11*)
- When children have mild-to-moderate hearing loss, they can usually manage



Figure 10: A girl hearing and repeating after her mum for the first time with a headset on



Figure 11: Learning to pronounce the letter “p” by blowing cotton balls

- in an ordinary school if they are given a hearing aid
- Teachers need to know how they should manage hearing impaired children
 - Place the child correctly at the front of the classroom to see and hear the teacher well
 - A teacher’s face should be well lit
 - Turn to the child when speaking
 - Do not speak when writing on the blackboard
 - Talk clearly and not too fast and repeat questions and answers from other pupils
 - Write important things down
- Assistive devices *e.g.* a hearing loop in the classroom helps children with hearing aids to hear the teacher’s voice directly in the ear

- When children have profound hearing loss or deafness they, as well as the whole family, need to learn sign language to communicate. The child also needs to be referred to a school for the deaf
- If a child has profound hearing loss and is diagnosed at an early age (before 2-3 years) a cochlear implant can provide useful hearing. After the age of 3 years a cochlear implant will give more limited hearing and permanently delayed speech and language development¹⁰. (See Chapter: [Cochlear Implants in Developing Countries](#))

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