HYPOACUSIS IN CHILDREN: hearing to grow up
INTRODUCTION

Methodology
International bibliographic resources were used to analyze a number of international clinical and laboratory studies involving the use of screening programs and hearing aids on children suffering from hearing loss.

Project Working Group
This review is the result of a study of the scientific literature available on the topic conducted by Prof. Edoardo Arslan, Audiology and Phoniatrics Department – University of Padua – Treviso Hospital, Prof. Dr. med. Annette Limberger, Aalen University of Applied Sciences, Aalen, Germany, Dr. Natalie Loundon, ENT Department – Prof. E. Garabedian équipe – Armand Trousseau Children’s Hospital – Paris.

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Most children with congenital hearing loss suffer from hearing impairment at birth which can usually be detected through newborn infant hearing screening. At least half of all cases of deafness and hearing impairment can be prevented through early detection. Otherwise, acoustic amplification or cochlear implants, supported by hearing and speech therapy, may be necessary.

Fitting, verifying and validating the adequacy of hearing aids is challenging and places great demands on the audiologist. The whole process has to be double checked to make sure that the child is provided with adequate amplification so that effective hearing can be restored through the appropriate therapy.

Early detection and quality therapy are the most powerful ways to optimize the impact of hearing rehabilitation on language development in children with profound congenital hearing loss: the use of increasingly sophisticated hearing aids or cochlear implants makes it possible to offset the hearing loss and recover the hearing ability needed for satisfactory language and speech perception development in almost all the instances involving children suffering from impaired hearing.

The purpose of this review is to provide useful information about hearing loss in children, diagnosis and possible solutions, in order to improve their future and health.

The Authors
01 Hearing loss in children: from early detection to treatment
Hearing loss in children: from early detection to treatment

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Key factors and global perspective
Hearing impairment in children worldwide constitutes a particularly serious obstacle to their optimal psychological development. Deafness can, in fact, affect language acquisition, learning ability and social integration. The cost of special education and lost employment due to hearing impairment can be a burden to a country’s economy. An estimated 1 to 2 out of every 1,000 newborns suffers from hearing impairment at birth which is detectable through newborn infant hearing screening. Some congenital hearing loss, however, may not become evident until later in childhood [Erenberg et al. 1999]. Severe, early deafness is present in 0.4 to 1.5 out every 1,000 children and at least 50% of these cases is attributable to genetic causes [Marazita et al. 1993].

There are two types of hearing loss. Conductive hearing loss is a problem in the outer or middle ear which affects a child’s quality of life, as in the case of middle ear problems. Typically conductive hearing loss is temporary, with slight to moderate hearing loss and can be treated either surgically or through drug therapy. Sensorineural hearing loss is a pathology of the inner ear, cochlea or hearing nerve; it is typically permanent and may require the use of a hearing aid or cochlear implantation (CI), along with hearing and speech therapy.

Congenital hearing loss can be hereditary or related to problems encountered during pregnancy and childbirth. The last position statement of the Joint Committee on Infant Hearing (2007) discusses the key risk indicators associated with permanent hearing loss in childhood. These include admission of the newborn to neonatal intensive care for more than 5 days or any of the following, regardless of the duration: extracorporeal membrane oxygenation (ECMO), assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix);
hyperbilirubinemia which requires a blood transfusion. Other risk indicators include any viruses a mother may have had during pregnancy, particularly during the first three months such as German measles, toxoplasmosis, and CMV [JCIH 2007 Pediatrics]. Acquired causes that can lead to hearing loss at any age, mostly in childhood, include infectious diseases like meningitis, measles, mumps and syphilis. Use of ototoxic drugs at any age, along with some antibiotic and anti-malarial drugs, can damage the inner ear. Head injury or injury to the ear can cause hearing impairment. Hearing impairment can be the cause of a heavy social and economic burden for individuals, families, communities and countries. Several socio-economic studies report [Abrams et al. 2002] that the social costs, in terms of rehabilitation and exclusion from a country’s socio-economic system, are in fact much higher when hearing impairment in children is not detected early and adequately treated.

Hearing impairment in children may delay development of language and cognitive skills, thus hindering progress in school. The extent of the delay depends on the degree of hearing loss. Later in life, hearing impairment often makes it difficult to obtain, perform, and keep jobs. Hearing-impaired children and adults are often stigmatized and socially isolated. Globally, the poor suffer more from hearing impairment because they cannot afford the preventive and routine care needed to avoid hearing loss. With limited or no access to ear and hearing care services, they are unable to obtain the hearing aids needed to make the disability manageable. Hearing impairment may also make it more difficult for these individuals to escape poverty or social isolation. At least half of all cases of deafness and hearing impairment are avoidable through early prevention. A large percentage can be treated through secondary preventive measures designed to reduce the hearing disability through early detection and suitable management as the correct rehabilitation can effectively restore hearing. With regard primarily to underdeveloped and some developing countries simple strategies for global prevention must include:

- improving prenatal and perinatal care, including promotion of safe deliveries;
- avoiding the use of ototoxic drugs, unless prescribed by a qualified physician and properly monitored for correct dosage;
- referring high risk babies, such as those with family history of deafness, those born with life-threatening conditions, for early hearing screening;
- assessment, diagnosis and treatment of hearing when required.

Conductive hearing impairment can be prevented by increasing the prevention of otitis in children with adequate specialized medical care and surgery. It should be noted, however, that the global production of hearing aids meets less than 10% of global demand. In developing countries, fewer than 1 out of 40 people who need a hearing aid have one [WHO 2012].

**New insights into auditory plasticity**

Early rehabilitation of a deaf child’s auditory function can help to correct the hearing deficit, as well as facilitate the maturation of the central auditory system and the auditory cortex as has been demonstrated in experiments involving deaf animals and humans. Patients with cochlear implants are useful in the study of hearing loss and help to understand how and when the brain can overcome the lack of auditory input. Electrophysiological studies in humans show maturation of evoked responses with cochlear implantation (CI) stimulation [Sharma et al., 2007]. However, a period of maximum receptiveness to auditory stimulation (sensitive period) occurs both in animals and in humans which is controlled by genetically determined processes and during which the principal changes and the organization of the neural pathways which are the foundation of hearing and language perception take place. If, during this period, the peripheral auditory input is not adequate the organization of the central hearing pathways will take place but in an incomplete manner. The absence of hearing experience during this sensitive period results in a substantial reduction of synaptic activity (computational power) of the cortex in deaf animals as compared with hearing animals. The auditory cortex, located in the temporal lobe (Figure 1) contains the majority of neurons present in our brains at the time of birth, but they have few and randomly distributed synaptic connections.
Synapses, at the basis of the neural circuits, develop very rapidly during the first two years of life and are tightly linked to the hearing experience. They then decrease from the third year of age on and constitute the base of the neural circuits devoted to hearing perception, specifically of language, which is the most important distinguishing characteristic of man’s ability to hear.

Figure 1
(taken from Kral A, O’Donoghue GM. 2010) Development of the synapses in the Central Auditory System and Auditory cortex. The postnatal development on the auditory cortex shows that the number of dendritic trees is highest at the age of 4 years in children with normal hearing (Huttenlocher and Dabholkar AS. 1997). Peak synaptic density has been observed at 2 to 4 years in children with normal hearing. Subsequently, synaptic counts decrease because unused synapses are eliminated. The same behaviour can be observed in congenitally deaf cats: the lack of auditory input in newborns, as shown in the lower part of the figure, has two main effects: retarded growth of synaptic circuits and a slight increase in the overall amount, probably explained by the permanence of non-functioning synaptic circuits.
Delaying effective auditory abilitation beyond this window markedly decreases brain adaptability and speech comprehension. This situation, called auditory deprivation, results in a lack of organization by the central auditory processor which affects the size and structure of the neural networks (they become smaller) and the synapses which becomes less reversible after the physiologic period of language development. In other words, the auditory input serves as a modulator and regulator of development of the central auditory processor which organizes itself around the acoustic information and neural impulses coming from the periphery. With regard, specifically to the auditory system, the lack of auditory stimulus will result in incomplete or inadequate development of the linguistic processor. The development of the human auditory system involves several sensitive periods relating to the auditory, phonetic and phonologic, syntactic, and semantic aspects of language. These periods probably reflect the maturation of the various cortical areas. Normal brain maturation also calls for the ability to respond through appropriate multimodal interaction, which is affected by deprivation. In persons who have become deaf after the acquisition of spoken language, brain activity triggered by a CI can be observed in non-auditory regions, with visual centres contributing to comprehension of speech through lip-reading. However, cross-modal reorganization can also cause deterioration of the auditory performance in persons and animals with congenital deafness if the critical period is exceeded. Based on these findings and language testing, effective hearing abilitation is recommended in the first 1 to 2 years of life, including CI surgery if needed. With the use of universal newborn hearing screening, this is feasible in the first year of life [Kral and O’Donoghue 2010].

Universal newborn hearing screening
The basic treatment for hearing impairment in children consists in early detection, along with the selection of adequate hearing aids and rehabilitation. Countries with different healthcare systems, as well as economic and social circumstances, have successfully implemented newborn and infant hearing screening programs. Although the aetiology of congenital or early-onset hearing loss most likely varies from country to country, there is widespread agreement that at least half of such hearing loss is due to genetic mutations. Viruses (such as cytomegalovirus, German measles, and meningitis); diseases (such as measles, mumps and chronic otitis media); adverse perinatal conditions (such as birth asphyxia, low birth weight and hyperbilirubinemia) and head trauma can also cause hearing loss. Regardless of its cause, unidentified hearing loss at birth or during the first few years of life adversely affects speech and language development, as well as success in school and social-emotional development. In the absence of universal hearing screening programs for newborns and infants, a significant number of cases of children with hearing loss are not detected until well beyond the critical period resulting in a state of irreversible hearing loss. In fact, it is not unusual for the diagnosis of milder hearing loss and unilateral hearing loss to be delayed until children are six years of age or older. When detection and intervention occur during the first few months of life, infants and young children with hearing loss perform dramatically better in scholastic activities including vocabulary development, articulation, social adjustment and behaviour.

Most existing newborn and infant hearing screening programs target permanent sensory or conductive hearing loss, averaging 30-40 dB or more in the frequency region important for speech recognition (approximately 500–4000 Hz). There is growing agreement that milder hearing loss (20–30 dB) is also important and needs to be detected and treated early because of the negative consequences of such loss on the later development of children. Some programs are designed to identify only bilateral hearing loss, but there is again growing agreement that the identification of unilateral hearing loss is also important and valuable. Similarly, fluctuating conductive hearing loss caused by otitis media are generally not targeted by newborn hearing screening programs, even though chronic otitis media has serious negative consequences.

A number of different approaches can be used to identify hearing loss in newborns and infants. There is widespread agreement that the best approach is the universal newborn screening procedure using otoacoustic emissions (OAE) testing or auditory brainstem response (ABR). However, where such programmes are not possible because of financial limitations, or because appropriate equipment and personnel are not available, or because of other constraints – other approaches can be valuable ad interim. These include family questionnaires or behavioural analyses. Such methods produce high levels of both false negatives and false positives with babies less than 12 months old. OAE or ABR measurements have been shown to be effective methods of screening for hearing loss in newborns and infants. OAEs measure the status of the peripheral auditory system extending to the cochlear outer hair cells. OAE measurements are obtained from the ear canal by using a sensitive microphone within a
probe that records cochlear responses to acoustic stimuli. The ABR measurements are obtained from surface electrodes that record neural activity generated in the auditory nerve and brainstem in response to acoustic stimuli delivered via an earphone. Screening ABR measurements are usually automated (AABR) and reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway.

The benefits of early hearing detection and intervention (EHDI) programs which facilitate early detection of and therapy for infant deafness are well known. It is only through these preventive steps and the subsequent therapy that today we are able to detect deafness in the first few months of life and subsequently program audiological and diagnostic procedures, as well as determine the medical treatment, surgery and devices needed while also helping to educate the children suffering from hearing impairment and their families. Successful newborn and infant hearing screening programs have been implemented in many different countries using a variety of screening methods and protocols, while working closely with the public healthcare agencies, social services and schools. Such programs are widely accepted as both highly worthwhile, with acceptable cost/benefit ratios and should, therefore, be offered in all countries. Although every nation should work to implement universal newborn hearing screening using OAE or AABR, interim approaches using targeted screening of children based on risk indicators or based on questionnaires can also be beneficial. Whatever approach is used, it is important that the EHDI program is linked to existing healthcare programs, as well as social and educational services, and that the procedures and outcomes of the program be documented so that ongoing quality control activities can be implemented and experiences shared [WHO 2009].

Early rehabilitation of hearing and language

Improved and more widespread screening programs make it possible to identify more infants with hearing loss at an increasingly younger age. Helping families find and access appropriate therapies for their infants is the key to the success of screening programs. Until the 1990s, children born with significant hearing loss typically were not identified until they reached 24-36 months of age. Better coordinated and responsive follow-up systems ensure earlier detection and treatment. Auditory brainstem response (ABR) and otoacoustic emissions (OAE) screening techniques paved the way for quick, effective ways to screen the hearing of babies. Healthy People 2000, a national health promotion and disease prevention initiative in the US, indicated that testing for congenital hearing loss should begin at 12 months of age. A decade later, Healthy People 2010 includes additional benchmarks: hearing screenings by one month, audiologic evaluation by three months, and early intervention by six months of age. If amplification is indicated for a baby with hearing loss, early, appropriate amplification requires accurate estimates of hearing sensitivity. For infants, a typical audiological test battery consists of ABR, tympanometry and acoustic reflex testing, and OAEs, with behavioural audiometric testing when developmentally feasible. Data about threshold agreement between ABR and behavioural audiometric tests for children have been published, supporting the use of both tools. Delays in finding the correct amplification may be due to problems with scheduling, the need for repeat tests, suspicion of auditory neuropathy/dys-synchrony, and the cost of hearing aids. Delays in fitting the hearing aid are also likely for babies who are medically fragile or very premature.

After the diagnostic procedure, the first step is to fit the child with a hearing aid. The process can be thought of as a five-step approach:

1. Selection
2. Fitting
3. Counselling
4. Fine-tuning
5. Follow-up

The selection stage coincides with the beginning of the process that will lead to the development of the infant’s communication skills. The family helps choose earmolds and hearing instruments; options are restricted by the relatively small size of infants’ ears. The earmold material should be a soft material, typically silicone or vinyl, which does not hurt the infant’s ears. Typically recommended pediatric hearing aids are binaural behind-the-ear and FM-compatible with flexible electroacoustic characteristics. Acoustic feedback occurs often with babies, as they quickly outgrow earmolds, but nowadays the digital hearing aids have feedback reduction circuits, which make maintenance of the earmold less critical.

In general there is some agreement, albeit from varying sources, as to the specific circuitry to be used with infants. Wide Dynamic Range Compression (WDRC)
Hearing aids are appropriate for children who have mild to moderately severe hearing loss, according to a review of the studies made by Palmer and Grimes (2005). They state that evidence supports the use of low compression thresholds, moderate compression ratios, and fast attack times.

There is also an approach developed by pediatricians designed to ensure that specified sensation levels for inputs and outputs in amplified speech are reached. The Desired Sensation Level (DSL) approach, developed by Seewald and colleagues, works with these targets.

Subsequent assessment should be based on the hearing sensitivity of the child wearing the hearing aid and the hearing aid’s electroacoustic inputs and determine whether speech is audible using a wide range of inputs. Electroacoustic assessment is frequently the only feasible option with very young babies. It provides frequency-specific information about the audibility of a wide variety of speech inputs, as well as estimating the individual ear amplification response (RESR).

Because infants are unlikely to tolerate the repeated use of probe-microphone devices, electroacoustic assessment for them includes real-ear-to-coupler-difference (RECD) measures combined with coupler values to predict the real-ear-aided-response (REAR) and RESR, taking into account the rapid physiological growth of the ear in babies.

To promote the consistent use of a hearing aid, parents and/or caregivers need information about proper care, the appearance and benefits of the devices. It is very important that parents are involved in the fitting of the hearing aid. Parents should be shown how to take care of the earmolds and hearing aids; insert the earmolds and put the hearing aids in place; as well as remove the devices. Daily sound checks are vital because infants cannot tell anyone when a hearing aid is malfunctioning. The family must be supplied with hearing aid maintenance tools. Child-friendly retention devices, including pediatric tone hooks, help babies accept and keep the devices. Many manufacturers provide hearing device care kits.

In order to assess the benefits of hearing aids in older children and adults speech perception is measured while the device is being worn. Babies require further assessment measures to be implemented, which often are subjective and depend on feedback from parents or the clinician’s observations. However, the ultimate assessment tool is the measurement of speech and language development. In the early months of life, babbling and phoneme development should be monitored carefully. Tests have been developed in many languages for testing speech perception abilities in children based on age, beginning at 6 months. The medical staff, parents, therapists and audiologists all must collaborate in order to determine if changes in amplification are needed.

Following the detection and confirmation that hearing loss exists, and subsequent to the fitting of the hearing aid, a reasonable follow-up schedule for children less than 2 years old includes check-ups every three months. At each check-up hearing is monitored, hearing aids are tested and adjusted, and new earmolds are made when needed. Check-ups every six months are appropriate for most children from the ages of 2 to 6, with yearly check-ups thereafter, unless risk factors indicating the hearing loss could worsen are present [Hoffman and Beauchaine 2007].

When well fitted hearing aids are still not enough to assist speech perception, CI should be considered. Several factors appear to influence the process involved in deciding whether or not a child should have a CI, including the family’s preferences, the child’s cognitive and neurological state, and the amount of the child’s residual hearing. Clinicians have, however, expressed concern about whether children with hearing loss more than 90 dB HL, average 500-1000-2000 Hz, were appropriate candidates for cochlear implantation. For thresholds ranging from 70 to 90 dB HL, borderline cases, therefore, clinicians were more likely to rely on additional information including the child’s spoken language, progress in treatment, social interaction, academic ability and classroom comprehension [Fitzpatrick et al. 2009].

In light of the ever changing landscape relative to CI eligibility, it is important to stay current on the technology and the criteria used to determine eligibility in order to be able to help families find the most appropriate solution for their children.

An increasing amount of scientific evidence points to the importance of implanting the devices at the right time in a child’s life, particularly those with greater amounts of residual hearing, as well as the importance of reducing hearing aid trial periods. Future studies will need to focus on providing the data needed to assist clinicians in helping families make the right decisions for their children [Tobey 2010].
To conclude, today it is possible to say that the condition of being deaf-mute, namely the most severe communication disorder, which occurred 20 years ago has finally disappeared.

Early Hearing Detection and Intervention programs based on universal hearing screenings, early detection and treatment are the key to diagnosing and treating hearing impairment in children (Figure 2).

![Figure 2](image1.png)

The three Components of Early Hearing Detection and Intervention Programs (from: http://www.jcih.org/).

The use of increasingly sophisticated hearing aids or cochlear implants, in the case of severe hearing loss (Figure 3), makes it possible to offset the hearing loss and recover the hearing ability needed for satisfactory language and speech perception development in almost all the instances involving children suffering from impaired hearing.

![Figure 3](image2.png)

Diagnostic and therapeutic flow from birth to hearing restoration.

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**FLOW CHART FROM SCREENING TO HEARING RESTORATION IN CHILDREN**

1. **SCREENING**
2. **EARLY AUDIOMETRIC MEASUREMENTS**
3. **FIRST ACOUSTIC AMPLIFICATION**
4. **BEST ACOUSTIC FITTING**
5. **HEARING AND LANGUAGE EVALUATION**
6. **HEARING AID**
7. **COCHLEAR IMPLANT**

To act against auditory deprivation
To maximize Central Auditory System input

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References


Children and hearing aids, hearing loss rehabilitation
Fitting children with hearing aids is quite challenging and the approach varies depending on the age of the child. In babies and toddlers the fitting process is based primarily on objective findings like ABR (auditory brainstem response), otoacoustic emissions (OAE) and impedance measurements. In older children, from age two and a half or school-age children, the fitting procedure may rely much more on subjective measurements like the VRA (visual reinforcement audiometry) and speech audiometry.

Fitting hearing aids in babies
In this age group the audiologist is dependent on ABR, otoacoustic emissions and impedance measurements. It is important, therefore, to use frequency dependent ABR measurements with both tone bursts or notched noise signals. The measurements should be conducted with insert earphones to measure each side separately from the other.

The DSL method described by Scollie et al. involves an instrument for fitting hearing aids in babies and children which is widely accepted and scientifically proven to be adequate. The method calls for amplification of low level sounds like whispering, wind rustling in trees etc. to make them audible. On the other hand, this method limits the output level of the hearing aid, so that loud sounds, i.e. crying, clapping hands, can never be too loud (Scollie et al., 2005).

Based on this fitting procedure, it is possible to use electroacoustic hearing threshold estimates particularly for infants and very young children. The formula for the hearing aid prescription, however, uses behavioral thresholds. The levels detected in the frequency-specific ABR measurements in dB nHL (normalized hearing level) must therefore be corrected to behavioral thresholds (dB HL); in infants with sensorineural hearing loss (SNHL) the behavioral thresholds are 10-20 dB higher than the ABR threshold estimates (Stapells, Gravel,
& Martin, 1995). The DSL research group has established a correction from nHL to eHL (estimated hearing level) (Table 1). It is also based on the equipment and the parameter settings, so each clinic has to determine their own correction factors for each frequency. After correcting the nHL levels, the values can be used for the calculation of the amplification (Scollie et al., 2005).

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Correction factor (substract from ABR nHL-thresholds)</td>
<td>-15</td>
<td>-10</td>
<td>-5</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 1: Corrections for frequency-specific ABR measures (nHL → eHL)

Another main part of the fitting process is the measurement of the RECD (real ear to coupler difference). This means that it is necessary to measure the real ear settings with the individual earmold because of the smaller ear canal volume in small children. The smaller volume leads to a higher sound pressure level (SPL) in the eardrum. This would lead to over amplification and could also harm the haircells in the inner ear. To avoid this type of over amplification it is essential to measure the RECD. It is enough to measure one ear if the impedance measurements are the same for both ears and if they look physiologically the same. This measurement is also used to convert the dB HL values in dB SPL values because the hearing instrument also works in dB SPL. With this measurement it is possible to convert the hearing aid amplification and output levels to estimated real-ear measurements and to simulate real ear verification to match the assessment targets. The variation in the RECD is very high so if it is possible to obtain a real measurement it should be done. The RECD should also be re-measured when the earmold or when the middle ear status changes, for example if the child has ventilation tubes or suffered from transitory middle ear otitis (Bagatto et al., 2006).

Selecting the hearing aid

For infants, BTE-style (behind the ear) hearing aids are standard, they should also be equipped with:

- Direct audio input
- Locking mechanism on the battery door and volume control for safety reasons
- Ability to deactivate advanced features - directional microphone - noise reduction - multiple memories
- Choice of bright colours
- Possibility of obtaining a substitute device in the case of an emergency
- Appropriate number of frequency channels to allow for frequency/output shaping
- Wide dynamic range compression (WDRC) to prevent discomfort due to loud noise and distortion

Advanced features are mostly unnecessary in infants or very young children, but as they grow older and in the presence of more challenging, noisier environments, it might be helpful to activate noise reduction or directional microphones in special situations (for example, during speech therapy).

Verification

“The responsible audiologist wants to know as much as possible about the levels of the amplified sound that hearing instruments deliver into the ears of infants and young children. To this end, the audiologist must apply comprehensive and evidence-based verification strategies that are compatible with the characteristics and capabilities of this unique population. This is because the long-term implication of the fitting decisions we make are simply too important.” (Richard Seewald)

After the audiological assessment, the RECD measurement and the selection of the hearing aid, it is essential to measure the amplification and the output levels to exclude over or under amplification. In adults it is recommended to measure the real ear aided response (REAR) with a probe microphone in the ear canal. In infants this procedure is difficult to conduct because they have to keep calm for several minutes and the measurement of the maximum pressure output (MPO) requires a high intensity of narrow band signals.
(90-100 dB SPL). This signal may be startling or uncomfortable to the child. The RECD converts 2-cc coupler measurements in the test box to predicted real-ear measurements. With the SPLogram there is an additional instrument to determine whether most sounds are audible for the child and fit the dynamic range. The SPLogram displays the threshold, the recommended targets for average speech and the maximum power output as a function of frequency. For modern hearing aids it is also essential to use a speech based test signal like the ISTS (international speech test signal).

Fitting the hearing aid
The younger the child the more we are dependent on the observations of the parents and other caregivers, therefore questionnaires are designed for parents and other caregivers, such as teachers. As a result it is more important to find out how the child uses what is heard in everyday situations and not only what a child hears. Families are the primary observers of the behavior of their infants and young children, thus it is very important to teach the families how to observe their child’s speech/language and auditory behavior with and without amplification (Tharpe & Ryan, 2011).

Management of toddlers and older children
The principles of hearing aid selection and amplification are more or less the same. The audiological assessment differs insofar as subjective measures become more important, such as VRA and speech audiometry. The more accurate the assessment of the child, especially with insert earphones, the better the understanding of the behavioral threshold and the calculation of the amplification and maximum power output. With the utterance of the first words and development of initial language skills it is also possible to perform speech tests according to the developmental age.

With the developing child additional features such as directional microphones and FM-systems may be used more often, but the audiologist has to make sure that the parents and other caregivers are familiar with the use of these and know to switch them off if they are not being used, e.g. during training sessions.

Follow-up
In the first stage of the fitting process it is mandatory to see the child almost monthly, especially in the first year of life. During this time the anatomy of the outer ear changes very quickly and therefore it is necessary to manufacture new earmolds every now and then. With every new earmold the acoustic coupling changes and a new RECD-measurement has to be performed. In the second year a three month interval is recommended and can be prolonged in the third year to a six month interval. It is also of utmost importance to evaluate the speech and language development regularly in order to understand if cochlear implantation is opportune and to intervent at an early stage.

Conclusion
Fitting, verifying and validating the adequacy of hearing aids is challenging and places great demands on the audiologist. The whole process has to be double checked to make sure that the child is provided with the adequate amplification.
References


The use of cochlear implants in infant deafness
The use of cochlear implants in infant deafness

Average to profound conditions of congenital hearing loss affect 0.5-1 out of every 1,000 newborn babies. In cases of severe to profound hearing loss, where the auditory system leaves no hope of good-quality aural rehabilitation, the question of a cochlear implant arises. This is an aural rehabilitation device that most often proves to be effective, but which requires a good understanding of when it should be used and its limitations. In fact, building a baseline aural-verbal system means the involvement of peripheral hearing and the development of sensory and motor areas, as well as cognitive factors. In children, cochlear implants must be part of a long-term plan, and take into account medical, psychological, social and rehabilitation components.

How the cochlear implant works
A cochlear implant is an implantable prosthesis comprising a removable part and an implanted part. The external portion consists of a microphone, a voice processor and an antenna; the internal portion comprises a receiver and a strip of electrodes. The encoded sound information is transmitted to the internal part via the antenna. The intra-cochlear electrodes are activated depending on the frequencies to be transmitted. The spiral ganglion cells are thus directly stimulated through the cochlea.

The auditory information is digitalized for frequency bands ranging from 250 to 8,000Hz. The signal can be processed in several ways, depending on whether one gives priority to speed of stimulation or to the number of channels activated simultaneously. The encoding strategies will depend on the number of functional electrodes and processors. Every implant has its own specific ergonomic and electronic features, but the results in terms of speech therapy are the same.
The choice of an implant will depend on what the team is used to and the specific needs of each patient. Surgical insertion of the internal part requires several hospitalization days. During surgery, the effect of the stimulation can be measured either by electric evoked response produced through the implant, or by telemetry. These tests are designed to check that the implant-nerve connection is working and provide an indication of the effective stimulation thresholds in the post-operative stage. The implant is activated a few days after surgery. Approximately ten adjustment visits will be required during the first year, then just once or twice a year after that.

**Indications**

The criteria for pediatric implants were the subject of an international consensus in 1995 and more recently discussed in a French nationwide recommendation by the National Health Authority (Haute Autorité de Santé - HAS) in 2007, updated in 2011 (www.has-sante.fr):

- profound bilateral hearing loss
- thresholds with hearing aid greater than or equal to 60 dB
- open set word (OSW) intelligibility scores of less than 50%.

These guidelines are further developed based on results reported and the experience acquired by the teams involved with cochlear implantation and rehabilitation. The implants were found to be called for primarily in the presence of severe bilateral or asymmetric hearing loss, as well as partial or fluctuating hearing loss.

**The implantation team**

The implantation team is multi-disciplinary and comprises, in addition to a technical platform that suits the child, the following specialists: an ENT (Ear, Nose, Throat) surgeon, a speech therapist and a psychologist.

The role of this team is to determine whether the implant gives hope for more improvement with respect to a conventional device, and whether there are any obvious contraindications (surgical problems, a progressive medical pathology, psychological problems, etc.)

The pre-implant survey is an especially important time. The rehabilitation team in charge of the child is encouraged to discuss this project and provide the family with all the information needed. Each case is discussed with the team before any decision is made.

During this time any medical conditions or progressive pathologies for which special monitoring is required (visual, neurological, vestibular or cardiac abnormalities) may also be ascertained.

**Pre-implant survey**

The parents take part in a medical information session during which they are told how the implant works. The limitations and surgical risks are also discussed. An audiogram, with and without the child wearing the high-powered device, is used to verify the appropriateness of the device. Both tonal and speech perception tests are conducted in silence and in noisy conditions.

A clinical ENT examination serves to eliminate any sources of infection, and in particular to guide the search for related pathologies in cases of syndromic and/or genetic hearing loss, if that has not already been done. Depending on each case, specialized pediatric examinations may be called for, such as an ophthalmological exam or a neuro-pediatric consultation. An ear scan is used to detect any malformations of the inner ear and to ascertain the surgical approach needed, and an MRI is used to explore the labyrinth, the inner ear channel and the cerebral parenchyma.

Speech therapy testing is essential as it makes it possible to assess the child’s communication skills and linguistic abilities. It also helps define a speech therapy plan together with the family.

A meeting with the psychologist serves to assess the child’s cognitive and psycho-affective development and to better understand the family’s expectations and motivations.

Before performing any implantation, it is essential that optimal rehabilitation care will be available and that an appropriate plan for schooling has been developed.

**Post-implantation**

Several days after surgery, the first adjustments to the implant begin, and it normally takes 2 months before effective auditory thresholds are reached. The child’s progress in speech therapy is assessed regularly, and technical monitoring of the device is carried out in tandem with this.
The implant team should have many years of experience observing children with implants. This experience allows them to make correct decisions and guarantee the harmonious development of each child’s language perception and use, as well as monitor the speech therapy program. In general the results are good even if they depend on variables which include the type of hearing loss, age at the time of implantation, medical history, educational program and socio-cultural environment, etc. 6-9.

Progress and results
Post-implant results will depend on a number of factors, but the key factor for congenitally deaf children is how early the surgery is performed. Neurological research has, in fact, shown that there is a critical period for the development of the senses in general, and of the central auditory system, in particular. Development of the hearing-speaking loop depends on the nature of this development. When a child with profound hearing loss is given an implant, the recordings of cortical response obtained during auditory stimulation seem to confirm that normalization of the tracings occurs if the surgery is performed before age 3 and no normalization after age 7 10.

Analysis of brain functions confirms the notion that there is an optimal period for rehabilitation of profound congenital bilateral hearing loss.

There are numerous clinical studies on the various perceptive, linguistic and educational aspects of the effect of implantation in only one ear. Improvement in perception scores is constant when measured with traditional indicators, even if to varying degrees.

The perception and oral language skills of a group of children with congenital hearing loss who have had their implant for at least 3 years were compared to those of children with profound hearing loss without implants of similar age. The results show a substantial difference between the two groups 11. Comparison of children with profound hearing loss and children with no loss show that language tends to develop at the same rate in both groups after implantation 12. Clinical findings confirm the existence of a critical period during which children with profound congenital hearing loss should receive implants, namely around 24 months. There is a negative correlation between OSW scores and age at implantation in children with congenital hearing loss who receive implants after 30 months 13. Watson et al. 14 studied 176 children after they received cochlear implants; 3 groups were set up based on age at implantation: prior to age 3 (G1), between ages 3 and 5 (G2) and after age 5 (G3). After 5 years, there was oral communication in 83% of the cases in G1, in 63.5% of G2 and in 45.1% of G3. For children given implants before 24 months, word intelligibility scores were in the normal range at age 5. Children given implants before 16 months showed they could develop a proto-language more rapidly than children with no hearing loss and could catch up to them, and their phonetic development subsequently followed comparable patterns 15–17. This language development also made it possible for children to develop reading and educational skills at the same rate as children with no hearing loss.

Geers et al. studied the reading abilities of 181 deaf children, aged 8 to 10 years, who had had implants for 4 to 6 years. More than 51% of them achieved a reading level comparable to normal children of the same age. When there were no disorders associated with their hearing loss, 2/3 of the children could follow a standard school curriculum 18–20.

Cochlear implants are an effective means of rehabilitation for perception and usually lead to the development of oral language in children with profound congenital hearing loss. If there are no related problems, and if treatment is given early, normal progress in school is feasible.

Special indications

Bilateral Implantation
Bilateral implantation may be offered to implant candidates in cases of severe to profound bilateral hearing loss. Unilateral implantation promotes the development of oral language skills in deaf children in most cases. The hypothetical value of bilateral implantation is to rehabilitate the binaural hearing function, which makes it easier to understand speech in a noisy environment and helps with sound localization, which is impossible in cases of unilateral rehabilitation.

The degree of aural comfort obtained with bilateral implantation, which reduces the number of situations where there is only partial reception, could have an effect on the quality of speech and language skills in the long term.

The benefits of bilateral implantation have been studied in adults who have experienced hearing loss and, more recently, in children with congenital hearing loss 4. In the case of acquired or secondary hearing loss, following a case of meningitis, for example, restoration of binaural abilities can be quite good. These results, however, cannot be directly applied to children with congenital hearing loss as they have had no prior hearing
experience and above all they have not developed binaural perception which depends on the development of the central hearing system. Gordon et al. 21–24 found evidence of incomplete processing of bilateral cortical responses in cases of late implantation of two separate devices which were implanted more than 2 years apart. These objective factors confirm the clinical findings that show better results in children given both implants before the age of 18 months. The most obvious effect of dual implantation to be highlighted is the improvement in hearing in noisy environments, which may range from 2 to 6 dB 25,26. It is possible that a certain degree of localization may be recovered, but this depends on prior hearing experience, age at implantation, and the time lapsed between the two implantations 27,28.

The effect of dual implantation on the development of speech is, however, less well known, due to the many factors involved in the acquisition of speech and to tiny differences that are hard to determine. Studies made of children who received dual implants before age 3, whether simultaneously or sequentially, show that when compared to peers with single implants there is a tendency for more natural usage and earlier attempts at oral communication 15,29.

In order to optimize the development of binaural hearing in a child suffering from profound hearing loss at birth, it seems important to offer dual implantation at a very early age.

The improvement in word recognition scores under noisy conditions may be as much as 50% in cases of bilateral rehabilitation. The effect of bilateral implantation on speech acquisition and on quality of life remains to be determined.

Cochlear Ossification
After a case of bacterial meningitis, hearing loss may occur due to incipient cochlear ossification, and may do so in the 2 years following the disease. Under such conditions, implantation becomes urgent, before complete ossification occurs. In cases of advanced ossification, certain surgical techniques may be employed (micro-drilling, bundles of electrodes), but the results continue to be disappointing 8,30–33.

Ear Malformations and Hypoplasia of the Auditory Nerves
In cases of cochlear malformations, a number of problems arise. The cochlear or cochleo-vestibular cavity must be large enough to admit a minimum number of electrodes and the cavity must be anatomically accessible. It is also necessary to ensure that the auditory nerve is present. In cases of complex malformations, the neural interface is not always functional. Surgical risks include geyser's, traumatic facial paralysis, migration of the electrode support and meningitis. Speech results are contingent on the type of malformation and on auditory experience 34,35.

Children without an auditory nerve detectable by MRI (Magnetic Resonance Imaging) are, in principle, not candidates for implantation. In some situations, there is a divergence between the clinical diagnosis and imaging results, which suggests that an underdeveloped nerve may be present but not visible. In that case, the possibility of an implantation may be discussed, but on average such children show perception results that are more limited than in other patients; so due care must be taken with indications 36–38.

Related Disabilities
Approximately 1/3 of deaf children also suffered from related disabilities or ones that will subsequently appear as the child develops. Some may suffer from visual deficiency, while others will have quite limited speech and may not be suitable for surgery (mental retardation, psychopathological disorders). Children with mental retardation may benefit from an implant in terms of hearing perception, but their linguistic development will be limited or non-existent 39,40.

It is, therefore, especially important to explain the limits and constraints of the implant to the family, and to ensure that appropriate therapy can be set up by the rehabilitation team through careful examination of each individual case 7,41.

Early Implantation
In order to intervene during the critical period, a decision must be made by 12 months of age. The hypothetical advantages of very early implantation must be weighed against problems specific to the newborn. It is indeed not always easy to confirm a diagnosis of profound hearing loss before 6 months of age, even using current objective methods and subjective examination. Furthermore, observation of the child’s development is limited and any associated pathology may be hard or impossible to detect. This may result in incorrect decisions or cause the parents’ expectations to be unrealistic. Lastly, one must be very familiar with how the infant copes with surgery and anesthesia, as well as the technical problems related to adjusting the implant. Treatment must be provided by an implantation unit with experience working with very young children. In this case, the risks related to anesthesia and surgery are comparable to those of children who receive implants after...
12 months (3.2% versus 2.3% - 4.1%) 42. In clinical terms, it is hard to detect any difference in development between the group of children implanted before 12 months and children implanted between 12 and 24 months. A recent review of the literature nevertheless found that 40% of infants showed improved scores in comparison to those given an implant between ages 12 and 24 months 43.

Residual Auditory Function
The idea of implantation in cases of residual audition is initially based on the possibility of preserving audition despite the insertion of the electrode support into the cochlea. Preserving audition requires a minute, flexible electrode design and a minimally invasive surgical technique. For a group of children and adults, Skarzynski et al. found a post-operative loss of less than 5 dB in more than half of the patients implanted 44–49. More recently, Brown et al. found partial auditory preservation in 90% of a pediatric group implanted with a long electrode 50. Dowell et al. 51,52 found an improvement in OSW (open set words) perception in 98% of cases in comparison to those using a hearing aid.

Comparative studies show that children with residual hearing function and implants produce better results than their peers with the same level of hearing loss who used hearing aids, and that they were no different from those with an average hearing loss 53.

Conclusion
In children with profound congenital hearing loss, even if positive results in terms of perception most often appear quite rapidly after the use of the implant, the development of language is variable. One quarter of the children show progress in language, but slower than that of other normal children of their age group, and some have very serious language problems. These results clearly illustrate the effectiveness of the implant in the treatment of profound hearing loss, even if we must consider that language development is affected by many other factors, especially cognitive ones. However, early implantation and quality speech therapy are the most powerful ways to optimize the impact of hearing rehabilitation on language development in children with profound congenital hearing loss. In post-verbal hearing loss, the results in terms of perception are excellent. The quality of implant technology has also caused this therapy to be used with children suffering from severe or partial hearing loss.
References


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