

## Others' Publications about EHDl: December 2019 through April 2020

The *Journal of Early Hearing Detection and Intervention (JEHDI)* focuses on improving Early Hearing Detection and Intervention (EHDl) systems by publishing peer-reviewed articles describing current research, evidence-based practice, and standards of care that are relevant for newborn and early childhood hearing screening, diagnosis, support, early intervention, the medical home, information management, financing, and quality improvement.

Even though *JEHDI* is the only journal that focuses exclusively on improving EHDl systems, many other journals include some articles relevant to *JEHDI*'s aim. To help *JEHDI* readers stay up-to-date about information in other journals about improving EHDl programs, we provide titles and abstracts of recent publications that are relevant to EHDl systems. Articles are listed in alphabetical order by the last name of the first author and titles of all articles are hyperlinked to the source.

The EHDl-relevant articles described in the following abstracts are from all over the world – demonstrating the global relevance of EHDl systems. Many of the following abstracts focus on screening and diagnosis, suggesting that the fundamentals of the EHDl system still need to be improved. Many other articles are breaking new ground and suggesting creative innovations. For example:

- DeForte et al. described an app called Hear Me Read (HMR) which uses enhanced digital stories as therapy tools for speech, language, and literacy for children with hearing loss. The study evaluated the user experience of the HMR app through a focus group study with caregivers and their children. The findings suggest that such educational apps can be valuable for those with hearing loss who are pursuing listening and spoken language as a communication outcome.
- Diener et al. surveyed 365 caregivers whose children were being seen in an otolaryngology clinic at a tertiary pediatric hospital about their knowledge of and attitudes toward congenital Cytomegalovirus (cCMV) and cCMV screening. Caregivers frequently were unaware of cCMV and its implications. Attitudes toward cCMV screening generally were positive. A majority wanted to know if their child had cCMV even if asymptomatic and were willing to pay \$20 for cCMV screening. The results suggested that education on epidemiology and impact of cCMV may benefit both prevention of infection and attitudes toward screening.
- Guo et al. evaluated the efficacy of concurrent hearing and genetic screening in a general 239,636 eligible infants. They found 548 infants with hearing loss based on the physiological hearing screening, 41 infants who passed the hearing screening but likely had hearing loss based on the genetic screen, and 570 infants at risk for ototoxicity which is undetectable by hearing screening. They concluded that genetic screening complements newborn hearing screening by improving the detection of infants at risk of hereditary hearing loss and ototoxicity, and by informing genotype-based clinical management for affected infants and their family members.
- Kruyt et al. evaluated the efficacy of Bone-Anchored Hearing implants (BAHIs) in children based on 20 articles published between 2000 and 2017, encompassing 952 children with implants. They concluded that BAHIs are a safe method for hearing rehabilitation in children, although large differences between studies are observed. The outcomes of new surgical techniques and implant designs in the pediatric population seem promising, but more research is needed before definitive conclusions can be drawn.
- Le et al. examined the health related quality of life (HRQoL) in young children with low language or congenital hearing loss. Based on a sample of 108 children in Australia, they found that children with low language and with hearing loss had lower HRQoL than children with normal language; the worst HRQoL was experienced by children with both. They concluded that children with low language and congenital hearing loss might benefit from interventions targeting overall health and well-being, not just their impairments.
- Rabiço-Costa et al. in a study conducted in Portugal assessed the incidence of hearing loss in 51 children after the exposure to platinum drugs used to treat central nervous system tumors. They found ototoxicity in 23.5% of the children. Even though the use of chemotherapy for such tumors has significantly improved cure and survival rates, the ototoxicity resulting from platinum-derived chemotherapy may accompany patients for the rest of their lives (see related article by van As et al.).

- Tarhun examined the smoking habits of family members of 75 children with serious otitis media (SOM) and 50 healthy controls. The correlation between SOM and passive smoke was statistically significantly positive. They concluded that the effect of passive smoking is a preventable and controllable risk factor in the etiology of the SOM.

Abstracts for all 100 articles are listed below.

Genes (Basel). 2020 Jan 27;11(2). pii: E132. doi: 10.3390/genes11020132.

**[Enhancing Genetic Medicine: Rapid and Cost-Effective Molecular Diagnosis for a \*GJB2\* Founder Mutation for Hearing Impairment in Ghana.](#)**

[Adadey SM](#), [Tingang Wonkam E](#), [Twumasi Aboagye E](#), [Quansah D](#), [Asante-Poku A](#), [Quaye O](#), [Amedofu GK](#), [Awandare GA](#), [Wonkam A](#).

**ABSTRACT:** In Ghana, gap-junction protein  $\beta$  2 (*GJB2*) variants account for about 25.9% of familial hearing impairment (HI) cases. The *GJB2*-p.Arg143Trp (NM\_004004.6:c.427C>T/OMIM: 121011.0009/rs80338948) variant remains the most frequent variant associated with congenital HI in Ghana, but has not yet been investigated in clinical practice. We therefore sought to design a rapid and cost-effective test to detect this variant. We sampled 20 hearing-impaired and 10 normal hearing family members from 8 families segregating autosomal recessive non syndromic HI. In addition, a total of 111 unrelated isolated individuals with HI were selected, as well as 50 normal hearing control participants. A restriction fragment length polymorphism (RFLP) test was designed, using the restriction enzyme NciI optimized and validated with Sanger sequencing, for rapid genotyping of the common *GJB2*-p.Arg143Trp variant. All hearing-impaired participants from 7/8 families were homozygous positive for the *GJB2*-p.Arg143Trp mutation using the NciI-RFLP test, which was confirmed with Sanger sequencing. The investigation of 111 individuals with isolated non-syndromic HI that were previously Sanger sequenced found that the sensitivity of the *GJB2*-p.Arg143Trp NciI-RFLP testing was 100%. All the 50 control subjects with normal hearing were found to be negative for the variant. Although the test is extremely valuable, it is not 100% specific because it cannot differentiate between other mutations at the recognition site of the restriction enzyme. The *GJB2*-p.Arg143Trp NciI-RFLP-based diagnostic test had a high sensitivity for genotyping the most common *GJB2* pathogenic and founder variant (p.Arg143Trp) within the Ghanaian populations. We recommend the adoption and implementation of this test for hearing impairment genetic clinical investigations to complement the newborn hearing screening

Iran J Child Neurol. 2020 Winter;14(1):21-30.

**[Well-Being and Coping Capacities of Adolescent Students with Hearing Loss in Mainstream Schools.](#)**

[Adibsereshki N](#), [Hatamizadeh N](#), [Sajedi F](#), [Kazemnejad A](#).

**OBJECTIVES:** Coping strategies used by adolescents has an important role in preventing or decreasing their stresses and also increasing their well-beings. This study aimed at evaluating the coping capacity and well-being of adolescent students with hearing loss in mainstream schools and also the correlations between their coping strategies and positive characteristics of well-being (engagement, perseverance, optimism, connectedness and happiness (EPOCH).

**MATERIALS & METHODS:** In this correlational study, 122 adolescent students with hearing loss were randomly selected from mainstream schools. Data collection was done by EPOCH Measure of Adolescent Well-Being and the Ways of Coping Questionnaire (WAYS). The Spearman correlation coefficient was used for determining the correlations between variables.

**RESULTS:** The mean scores of using different coping strategies varied from 1.36 in problem solving to 1.44 in seeking support. Among the positive characteristics of well-being, happiness had the lowest (11.04) and connectedness showed the highest score (12.33). The findings also showed a significant correlation between all coping strategies and EPOCH, however there was a strong positive correlation between total coping strategy score and perseverance (0.648) and happiness (0.629).

**CONCLUSION:** Based on the results, the score of happiness in students with hearing loss was the lowest among positive characteristics of well-being and also happiness showed a strong association with total scores in coping strategies. Accordingly, interventional studies are needed to examine whether training students with hearing loss to use coping strategies is effective in increasing their happiness and overall well-being.

BMC Pediatr. 2020 Apr 20;20(1):175. doi: 10.1186/s12887-020-02080-2.

**[Parental knowledge and attitudes to childhood hearing loss and hearing services in Qassim, Saudi Arabia.](#)**

[Alsudays AM](#), [Alharbi AA](#), [Althunayyan FS](#), [Alsudays AA](#), [Alanazy SM](#), [Al-Wutay O](#), [Alenezi MM](#).

**BACKGROUND:** Successful audiology service delivery depends on support from the community, and agreement to utilize hearing healthcare programs. Assessment of parents' awareness regarding hearing loss (HL) and audiology services is necessary for the development of suitable hearing programs for children. Previous studies reported that early detection and intervention for hearing problems are typically strongly supported by parents. The current study sought to evaluate parents' knowledge and attitudes regarding childhood HL and hearing services.

**METHODS:** A cross-sectional study conducted at five centers in Qassim region of Saudi Arabia. A self-report questionnaire was administered to collect demographic data in addition to 31 questions regarding the knowledge and attitudes of parents toward HL. IBM SPSS Statistics for Windows, Version 21 was used for data analysis. A p-value cut-off point of 0.05 at 95% CI was used to determine statistical significance. The analyses examined the association between socio-demographic characteristics and knowledge and attitudes toward HL using chi-square tests.

**RESULTS:**

Overall, participants included in this study were 243 participants. Of these, 105 (43.2%) were fathers, and 138 (56.8%) were mothers. Ages ranged from 21 to 60+ years. Assessment of the prevalence of various aspects of knowledge and attitudes among parents toward childhood HL revealed that 103 participants (42.4%) possessed good knowledge, while 140 participants (57.6%) possessed poor knowledge. In contrast, the attitude analysis revealed that 224 participants (92.2%) expressed positive attitudes, while only 19 participants (07.8%) showed a negative attitude regarding audiology services. We found a significant association between age group and knowledge ( $p=0.002$ ).

**CONCLUSION:** Most parents in our sample possessed poor knowledge regarding childhood HL. However, most parents expressed positive attitudes regarding audiology services. The current findings suggest a need to increase awareness among parents regarding childhood HL.

Indian J Pediatr. 2020 Apr 13. doi: 10.1007/s12098-020-03260-9

**[Congenital Cytomegalovirus and Zika Infections.](#)**

[Angueyra C, Abou Hatab H, Pathak A.](#)

**ABSTRACT:** Congenital infections affecting newborn infants can have potentially devastating clinical outcomes. They are usually caused by viruses that infect mothers during pregnancy and are transmitted to the fetus or newborn during the prenatal, perinatal or postnatal periods. Congenital cytomegalovirus (cCMV) is the most common congenital infection affecting up to 2.5% of all live births. Even though most infected infants are asymptomatic at birth, cCMV is an important cause of neurodevelopmental impairment and represents the main cause of non-hereditary sensorineural hearing loss. Also, congenital Zika infection has emerged in recent years as a cause of microcephaly and neurodevelopmental delays. Currently, universal screening is not recommended for either infection in pregnant women or newborn infants. Therefore, screening for both conditions is based on multiple factors such as maternal immune status, exposure, and clinical manifestations of the infant. Use of antiviral medications on symptomatic cCMV has shown improvement in outcomes, in contrast with congenital Zika for which there are no therapeutic options available. Even though both viruses can be present in breast milk, there are no recommendations against breastfeeding in full-term infants. Close follow-up for affected infants is necessary to monitor for developmental delays and sensory impairments to implement interventional therapies at the earliest time possible.

Lang Speech Hear Serv Sch. 2020 Jan 8;51(1):68-73. doi: 10.1044/2019\_LSHSS-OCHL-19-0025.

**[Audiological Considerations for Managing Mild Bilateral or Unilateral Hearing Loss in Infants and Young Children.](#)**

[Bagatto M.](#)

**PURPOSE:** This clinical focus article describes considerations for recommending assistive hearing technology to infants and young children who have mild bilateral or unilateral hearing loss. These conditions present special challenges compared to bilateral permanent hearing losses that are moderate to profound in their degree in that the recommendation to proceed with technology is not as clear.

**CONCLUSION:** Current clinical practice guidelines and protocols for pediatric hearing aid fitting recommend managing these conditions on a case-by-case basis. Descriptions of key considerations for recommending assistive hearing technology for infants and young children with mild bilateral hearing loss or unilateral hearing loss are offered herein.

Ear Hear. 2020 Mar 6. doi: 10.1097/AUD.0000000000000862.

**[Perception of Child-Directed Versus Adult-Directed Emotional Speech in Pediatric Cochlear Implant Users.](#)**

[Barrett KC](#), [Chatterjee M](#), [Caldwell MT](#), [Deroche MLD](#), [Jiradejvong P](#), [Kulkarni AM](#), [Limb CJ](#).

**OBJECTIVES:** Cochlear implants (CIs) are remarkable in allowing individuals with severe to profound hearing loss to perceive speech. Despite these gains in speech understanding, however, CI users often struggle to perceive elements such as vocal emotion and prosody, as CIs are unable to transmit the spectro-temporal detail needed to decode affective cues. This issue becomes particularly important for children with CIs, but little is known about their emotional development. In a previous study, pediatric CI users showed deficits in voice emotion recognition with child-directed stimuli featuring exaggerated prosody. However, the large intersubject variability and differential developmental trajectory known in this population incited us to question the extent to which exaggerated prosody would facilitate performance in this task. Thus, the authors revisited the question with both adult-directed and child-directed stimuli.

**DESIGN:** Vocal emotion recognition was measured using both child-directed (CDS) and adult-directed (ADS) speech conditions. Pediatric CI users, aged 7-19 years old, with no cognitive or visual impairments and who communicated through oral communication with English as the primary language participated in the experiment ( $n = 27$ ). Stimuli comprised 12 sentences selected from the HINT database. The sentences were spoken by male and female talkers in a CDS or ADS manner, in each of the five target emotions (happy, sad, neutral, scared, and angry). The chosen sentences were semantically emotion-neutral. Percent correct emotion recognition scores were analyzed for each participant in each condition (CDS vs. ADS). Children also completed cognitive tests of nonverbal IQ and receptive vocabulary, while parents completed questionnaires of CI and hearing history. It was predicted that the reduced prosodic variations found in the ADS condition would result in lower vocal emotion recognition scores compared with the CDS condition. Moreover, it was hypothesized that cognitive factors, perceptual sensitivity to complex pitch changes, and elements of each child's hearing history may serve as predictors of performance on vocal emotion recognition.

**RESULTS:** Consistent with our hypothesis, pediatric CI users scored higher on CDS compared with ADS speech stimuli, suggesting that speaking with an exaggerated prosody-akin to "motherese"-may be a viable way to convey emotional content. Significant talker effects were also observed in that higher scores were found for the female talker for both conditions. Multiple regression analysis showed that nonverbal IQ was a significant predictor of CDS emotion recognition scores while Years using CI was a significant predictor of ADS scores. Confusion matrix analyses revealed a dependence of results on specific emotions; for the CDS condition's female talker, participants had high sensitivity ( $d'$  scores) to happy and low sensitivity to the neutral sentences while for the ADS condition, low sensitivity was found for the scared sentences.

**CONCLUSIONS:** In general, participants had higher vocal emotion recognition to the CDS condition which also had more variability in pitch and intensity and thus more exaggerated prosody, in comparison to the ADS condition. Results suggest that pediatric CI users struggle with vocal emotion perception in general, particularly to adult-directed speech. The authors believe these results have broad implications for understanding how CI users perceive emotions both from an auditory communication standpoint and a socio-developmental perspective.

Int J Pediatr Otorhinolaryngol. 2020 Mar 24;134:110017. doi: 10.1016/j.ijporl.2020.110017.

### [Audiologic testing in children with Down Syndrome: Are current guidelines optimal?](#)

[Basonbul RA](#), [Ronner EA](#), [Rong A](#), [Rong G](#), [Cohen MS](#).

**INTRODUCTION:** Down Syndrome (DS) is a Tier 1 risk factor for hearing loss. Guidelines exist to ensure close monitoring of children with DS for hearing loss. It is important to consider the timing of testing in order to obtain meaningful audiologic data in this high-risk population. The purpose of this study is to present hearing outcomes for children with DS during the first 8 years of life and to assess these outcomes in the context of current screening guidelines.

**METHODS:** Retrospective review of audiometric outcomes was conducted for children with DS age 8 or younger who presented to a multidisciplinary DS clinic between January 2014 to June 2017. Age at the time of testing, as well as test success rate and hearing loss type and severity were noted.

**RESULTS:** 131 patients were included in the study, 52% of which were male. 36% of the patients failed their newborn hearing screening and only 9% of those subjects had normal hearing on subsequent testing. Most hearing loss identified was mild and conductive in nature. Inconclusive results were most likely to be obtained at 6-10 months of age.

**CONCLUSION:** Hearing loss is common among children with DS. To optimize the quality of testing and avoid the need for sedation in followup testing, routine follow-up hearing screening should be performed either before 6 months of age or after 10 months of age.

Fetal Pediatr Pathol. 2020 Jan 25:1-10. doi: 10.1080/15513815.2019.1710788.

### [Negative Effects of Noise on NICU Graduates' Cochlear Functions.](#)

[Beken S](#), [Önal E](#), [Gündüz B](#), [Çakir U](#), [Karagöz İ](#), [Kemaloğlu YK](#).

**AIM:** To evaluate the adverse effects of noise on hearing.

**METHODS:** Thirty-two infants that had been admitted to neonatal intensive care unit (NICU) and 25 healthy controls were included in this study. Noise levels were recorded continuously during the hospitalization period.

**RESULTS:** All healthy controls passed the hearing screening tests before discharge and on the sixth-month follow up. Hospitalized infants had lower “Distortion Product Otoacoustic Emission Signal Noise Ratio” (DPOAE SNR) amplitudes (dB) at five frequencies (1001, 1501, 3003, 4004, 6006 Hz in both ears). DPOAE fail rates at 1001 Hz and 1501 Hz were higher than in hospitalized infants (81.8% and 50.0% vs 20.0% and 4.0%). Infants who failed the test at 1001 and 1501 Hz were exposed to noise above the recommended maximum level for longer periods of time.

**CONCLUSION:** Hearing tests performed at sixth-months of life were adversely affected in NICU graduates.

J Speech Lang Hear Res. 2020 Jan 15;63(1):321-333. doi: 10.1044/2019\_JSLHR-19-00230.

## [Associations Between Parenting Stress, Language Comprehension, and Inhibitory Control in Children With Hearing Loss.](#)

[Blank A](#), [Frush Holt R](#), [Pisoni DB](#), [Kronenberger WG](#).

**PURPOSE:** Parenting stress has been studied as a potential predictor of developmental outcomes in children with normal hearing and children who are deaf and hard of hearing. However, it is unclear how parenting stress might underlie at-risk spoken language and neurocognitive outcomes in this clinical pediatric population. We investigated parenting stress levels and the shared relations between parenting stress, language comprehension, and inhibitory control skills in children with and without hearing loss (HL) using a cross-sectional design.

**METHOD:** Families of children with HL ( $n = 39$ ) and with normal hearing ( $n = 41$ ) were tested. Children completed an age-appropriate version of the Concepts & Following Directions subtest of the Clinical Evaluation of Language Fundamentals and the NIH Toolbox Flanker Test of Attention and Inhibitory control. Caregivers completed the Parenting Stress Index-Short Form 4.

**RESULTS:** Parenting stress levels were not significantly different between parents of children with and without HL. A significant negative association was observed between parenting stress and our measure of language comprehension in children with HL. A negative association between parenting stress and inhibitory control skills was also found in families of children with HL, but not hearing children. The parenting stress-inhibitory control relationship was indirectly accounted for by delayed language comprehension skills in children with HL.

**CONCLUSION:** Even at moderate levels of parenting stress similar to parents of children with normal hearing, increases in parenting stress were associated with lower scores on our measures of language comprehension and inhibitory control in children with HL. Thus, parenting stress may underlie some of the variability in at-risk pediatric HL outcomes.

Int J Pediatr Otorhinolaryngol. 2020 Apr 8;134:110039. doi: 10.1016/j.ijporl.2020.110039.

## [Implementation of a neonatal hearing screening programme in three provinces in Albania.](#)

[Bussé AM](#), [Qirjazi B](#), [Goedegebure A](#), [Toll M](#), [Hoeve HL](#), [Toçi E](#), [Roshi E](#), [Carr G](#), [Simonsz HJ](#).

**OBJECTIVES:** The EUSCREEN study compares the cost-effectiveness of paediatric hearing screening programmes and aims to develop a cost-effectiveness model for this purpose. Alongside and informed by the development of the model, neonatal hearing screening (NHS) is implemented in Albania. We report on the first year.

**METHODS:** An implementation plan was made addressing objectives, target population, screening protocol, screener training, screening devices, care pathways and follow up. NHS started January 1st, 2018 in four maternity hospitals: two in Tirana, one in Pogradec and one in Kukës, representing both urban and rural areas. OAE-OAE-aABR was used to screen well infants in maternity hospitals, whereas aABR-aABR was used in neonatal intensive care units and in mountainous Kukës for all infants. Screeners' uptake and attitudes towards screening and quality of screening were assessed by distributing questionnaires and visiting the maternity hospitals. The result of screening, diagnostics, follow up and entry into early intervention were registered in a database and monitored.

**RESULTS:** Screeners were keen to improve their skills in screening and considered NHS valuable for Albanian health care. The number of “fail” outcomes after the first screen was high initially but decreased to less than 10% after eight months. In 2018, 11,507 infants were born in the four participating maternity hospitals, 10,925 (94.9%) of whom were screened in the first step. For 486 infants the result of screening was not registered. For the first screen, ten parents declined, eight infants died and one infant was discharged before screening could be performed. In 1115 (10.2%) infants the test either could not be performed or the threshold was not reached; 361 (32.4%) of these did not attend the second screen. For the third screen 31 (34.4%) out of 90 did not attend. Reasons given were: parents declined (124), lived too far from screening location (95), their infant died (11), had other health issues (7), or was screened in private clinic (17), no reason given (138).

**CONCLUSIONS:** Implementation of NHS in Albania is feasible despite continuing challenges. Acceptance was high for the first screen. However, 32.4% of 1115 infants did not attend the second screen, after a “fail” outcome for the first test.

### [Risk factors for permanent childhood hearing impairment.](#)

[Butcher E](#), [Dezateux C](#), [Knowles RL](#).

**OBJECTIVE:** While several perinatal risk factors for permanent childhood hearing impairment (PCHI) are known, association with gestational length remains unclear. We hypothesised that shorter gestational length predicts higher PCHI risk.

**DESIGN:** 19 504 participants from the UK Millennium Cohort Study (born 2000-2002, prior to newborn screening).

**METHODS:** Multivariable discrete-time survival analysis to examine associations between parent-reported PCHI by age 11 years and gestational length, plus other prespecified factors.

**RESULTS:** PCHI affected 2.1 per 1000 children (95% CI 1.5 to 3.0) by age 11; however, gestational length did not predict PCHI risk (HR, 95% CI 1.00, 0.98 to 1.03 per day increase). Risk was increased in those with neonatal illness, with or without admission to neonatal care (6.33, 2.27 to 17.63 and 2.62, 1.15 to 5.97, respectively), of Bangladeshi or Pakistani ethnicity (2.78, 1.06 to 7.31) or born to younger mothers (0.92, 0.87 to 0.97 per year).

**CONCLUSION:** Neonatal illness, rather than gestational length, predicts PCHI risk. Further research should explore associations with ethnicity.

### [Etiological profile of hearing loss amongst Lithuanian pediatric cochlear implant users.](#)

[Byckova J](#), [Mikstiene V](#), [Kiveryte S](#), [Mickeviciene V](#), [Gromova M](#), [Cernyte G](#), [Mataityte-Dirziene J](#), [Stumbrys D](#), [Utkus A](#), [Lesinskas E](#).

**INTRODUCTION:** Congenital sensorineural hearing loss is a heterogeneous disorder; its etiological profile varies between populations. Pathogenic variants of GJB2 gene are the major cause of non-syndromic hearing loss. Congenital cytomegalovirus infection (cCMV) is the most important prenatal etiological factor causing hearing loss and other disorders. Perinatal events, syndromes, postnatal infections or traumas are less common. Causes of the remaining one third of hearing loss cases are unknown.

**OBJECTIVES:** To determine the etiological profile of hearing loss in pediatric cochlear implant users in Lithuanian population.

**METHODS:** The data of 122 children (70 male/52 female; aged  $7.6 \pm 3.3$  years) cochlear implant users were analysed. Medical records of all children recruited in Santaros Clinics (Vilnius, Lithuania) were analysed to identify prenatal, perinatal, or postnatal risk factors based on the adapted list proposed by the Joint Committee of Infant Hearing. Genetic counselling and testing according to the scheme were performed to 101 children. DNA of 117 children was extracted from the DBS on Guthrie cards and CMV DNA detected using real time PCR.

**RESULTS:** Non-syndromic hearing loss was diagnosed in 65 cases (53.3%), 58 of which were GJB2 gene-associated; syndromic hearing loss was diagnosed to 8 children (6.6%). Perinatal (prematurity, low birth weight, hypoxia, hyperbilirubinemia, sepsis, ototoxicity, and meningitis) and postnatal (meningitis) risk factors were associated with hearing loss in 16 (13.1%) and 4 (3.3%) study participants respectively. CMV DNA was detected in 12 samples (9.8%). The cause of hearing loss remained unknown only for 17 (13.9%) children.

**CONCLUSIONS:** The major cause of HL in the current study was GJB2 gene alterations. Only 14% of the cohort had congenital hearing loss of unknown origin.

### [Deafness Gene Mutations in Newborns in the Foshan Area of South China With Bloodspot-Based Genetic Screening Tests.](#)

[Cao S](#), [Sha Y](#), [Ke P](#), [Li T](#), [Yuan W](#), [Huang X](#).

**PURPOSE:** The aim of this study was to determine the rate of deafness gene mutations in the Foshan area of South China.

**METHOD:** We enrolled the infants delivered in Foshan Maternity and Children's Healthcare Hospital. Deafness gene mutation was detected by HibriMax method. Our study tested 47,538 newborns within 3 days after birth, including 13 sites in four genes: *GJB2* (c.35 del G, c.176 del 16, c.235 del C, c.299 del AT, c.155 del TCTG), *GJB3* (c.583 C>T), *SLC26A4* (c.2168 A>G, c.919-2 A>G, c.1299 C>T), and *mtDNA 12S rRNA* (m.1555 A>G, m.1494 C>T, m.12201 T>C, m.7445 A>G). The birth condition of infants was collected, including sex, low or high birth weight, twins, and premature delivery.

**RESULTS:** In a total of 47,538 newborns, 1,415 were positively identified with deafness gene mutations. The total rate of the deafness gene mutation was 2.976%. The carrier rates of *GJB2* (c.35 del G, c.176 del 16, c.235 del C, c.299 del AT, c.155 del TCTG), *GJB3* (c.583 C>T), *SLC26A4* (c.2168 A>G, c.919-2 A>G, c.1299 C>T), and *mtDNA 12S rRNA* (m.1555 A>G, m.1494 C>T, m.12201 T>C, m.7445 A>G) mutations were 0.000%, 0.048%, 1.422%, 0.185%, 0.000%, 0.076%, 0.116%, 0.755%, 0.160%, 0.187%, 0.021%, 0.000%, and 0.006%, respectively.

**CONCLUSIONS:** Our study showed that the c.235 del C *GJB2* mutation was the leading deafness-related mutation in the Foshan area of South China. Deafness gene mutations screening in newborns detected by bloodspot-based genetic screening tests can help the diagnosis of newborn congenital hearing loss.

Int J Environ Res Public Health. 2020 Mar 17;17(6). pii: E1969. doi: 10.3390/ijerph17061969.

### [Increased Risk of Sensorineural Hearing Loss as a Result of Exposure to Air Pollution.](#)

[Chang KH](#), [Tsai SC](#), [Lee CY](#), [Chou RH](#), [Fan HC](#), [Lin FC](#), [Lin CL](#), [Hsu YC](#).

**ABSTRACT:** Whether exposure to air pollution is associated with developing sensorineural hearing loss (SHL) remains controversial. Using data from the National Health Insurance Research Database, we recruited a total of 75,767 subjects aged older than 20 years with no history of SHL from 1998 to 2010, and they were followed up until SHL was observed, they withdrew from the National Health Insurance program, or the study ended. The subjects were evenly exposed to low-level, mid-level, and high-level carbon monoxide (CO) and nitrogen dioxide (NO<sub>2</sub>). The incidence rate ratio of SHL for patients exposed to high-level CO was 1.24 (95% confidence interval (CI) = 1.14-1.36). The NO<sub>2</sub> pollutants increased the incidence rate ratios of SHL in mid-level NO<sub>2</sub> and high-level NO<sub>2</sub> exposures by 1.10 (95% CI = 1.10-1.32) and 1.36 (95% CI = 1.24-1.49) times, respectively. The adjusted hazard ratio (adj. HR) of SHL in patients exposed to high-level CO was 1.45 (95% CI = 1.31-1.59), relative to that of patients exposed to low-level CO. Compared to patients exposed to low-level NO<sub>2</sub>, patients exposed to mid-level NO<sub>2</sub> (adj. HR = 1.40, 95% CI = 1.27-1.54) and high-level NO<sub>2</sub> (adj. HR = 1.63, 95% CI = 1.48-1.81) had a higher risk of developing SHL. The increased risk of SHL following the increased concentrations of air pollutants (CO and NO<sub>2</sub>) was statistically significant in this study. In conclusion, the subjects' exposure to air pollution exhibited a significantly higher risk of developing SHL in Taiwan.

Mol Biol Rep. 2020 Apr 22. doi: 10.1007/s11033-020-05460-0.

### [Mesenchymal stem cells for sensorineural hearing loss: a systematic review of preclinical studies.](#)

[Chorath K](#), [Willis M](#), [Morton-Gonzaba N](#), [Moreira A](#).

**ABSTRACT:** Sensorineural hearing loss (SNHL) is the most common form of hearing loss that is routinely treated with hearing aids or cochlear implants. Advances in regenerative medicine have now led to animal studies examining the possibility of restoring injured hair cells with mesenchymal stem/stromal cell (MSC) administration. We conducted a systematic review and meta-analysis to collate the existing preclinical literature evaluating MSCs as a treatment for SNHL and quantify the effect of MSCs on functional hearing. Our protocol was published online on CAMARADES. Searches were conducted in four medical databases by two independent investigators. Twelve studies met inclusion and were evaluated for risk of bias using SYRCLE. Rodent models were commonly used (n=8, 66%), while auditory brainstem response (ABR) and distortion product otoacoustic emissions (DPOAE) were the most frequent measures assessing hearing loss. MSCs were derived from multiple tissue sources, including bone marrow, adipose tissue, and umbilical cord blood and the dose ranged from  $4 \times 10^3$  to  $1 \times 10^7$  cells. Treatment with MSCs resulted in an improvement in ABR and DPOAE (mean difference -15.22, +9.10, respectively). Despite high heterogeneity and multiple "unclear" domains in the risk of bias, this review provides evidence that MSCs may have a beneficial effect in hearing function.

Pediatr Infect Dis J. 2020 Apr;39(4):273-276. doi: 10.1097/INF.0000000000002564.

### [Middle Ear Effusion in Children With Congenital Cytomegalovirus Infection.](#)

[Chung W](#), [Leung J](#), [Lanzieri TM](#), [Blum P](#), [Demmler-Harrison G](#), [Ahmed S](#), [Baer H](#), [Bhatt AR](#), [Brown F](#), [Catlin F](#), [Caviness AC](#), [Coats DK](#), [Edmonds JC](#), [Flores M](#), [Franklin D](#), [Gandaria C](#), [Greer J](#), [Griesser C](#), [Hussein MA](#), [Iovino I](#), [Istas A](#), [Jin HD](#), [Kelinske MK](#), [Klingen JT](#), [Laurente A](#), [Littman T](#), [Murphy M](#), [Miller J](#), [Nelson C](#), [Noyola D](#), [Paysse EA](#), [Percy A](#), [Reis S](#), [Reynolds A](#), [Rozelle J](#), [Smith O](#), [Steinkuller P](#), [Turcich M](#), [Vinson SS](#), [Voigt RG](#), [Walmus B](#), [Williams J](#), [Williamson D](#), [Yen KG](#), [Yow MD](#); [Congenital Cytomegalovirus Longitudinal Study Group](#); [Congenital Cytomegalovirus Longitudinal Study Group](#).

**BACKGROUND:** Sensorineural hearing loss (SNHL) is well described in children with congenital cytomegalovirus (CMV) infection, but limited data are available on middle ear effusion (MEE) occurrence in this population. We assessed the prevalence of MEE and the degree of transient hearing change associated with MEE among children with congenital CMV infection.

**METHODS:** Children with congenital CMV infection enrolled in a longitudinal study received hearing and tympanometric testing during scheduled follow-up visits annually up to 6 years of age. We used a generalized linear mixed-effect logistic regression model to compare the odds of MEE, defined as type B tympanogram (normal ear canal volume with little tympanic membrane movement) among patients categorized as symptomatic or asymptomatic based on the presence of congenital CMV-associated signs in the newborn period.

**RESULTS:** Forty-four (61%) of 72 symptomatic and 24 (28%) of 87 asymptomatic patients had  $\geq 1$  visit with MEE. After controlling for the number of visits, symptomatic patients had significantly higher odds of MEE (odds ratio: 2.09; 95% confidence interval: 1.39-3.14) than asymptomatic patients. Transient hearing decrease associated with a type B tympanogram ranged from 10 to 40 dB, as measured by audiometric air-bone gap in 11 patients. **CONCLUSIONS:** Among children with congenital CMV, MEE can result in transient hearing decrease, which can reduce the efficacy of a hearing aid in those with SNHL. It is warranted that children with congenital CMV infection and SNHL receive routine audiologic and tympanometric testing to better manage hearing aid amplification levels.

Otol Neurotol. 2019 Dec;40(10):1278-1286. doi: 10.1097/MAO.0000000000002410.

## **Long-term Outcomes in Down Syndrome Children After Cochlear Implantation: Particular Issues and Considerations.**

**Clarós P, Remjasz A, Clarós-Pujol A, Pujol C, Clarós A, Wiatrow A.**

**OBJECTIVE:** The aim of the study was to analyze the long-term outcomes after cochlear implantation in deaf children with Down syndrome (DS) regarding age at the first implantation and refer the results to preoperative radiological findings as well as postoperative auditory and speech performance. Additionally, the influence of the age at implantation and duration of CI use on postoperative hearing and language skills were closely analyzed in children with DS.

**STUDY DESIGN:** Retrospective analysis.

**SETTING:** Referral center (Cochlear Implant Center).

**MATERIALS AND METHODS:** Nine children with Down syndrome were compared with 220 pediatric patients without additional mental disorders or genetic mutations. Patients were divided into four categories depending on the age of the first implantation: CAT1 (0-3 yr), CAT2 (4-5 yr), CAT3 (6-7 yr), and CAT4 (8-17 yr). The auditory performance was assessed with the meaningful auditory integration scales (MAIS) and categories of auditory performance (CAP) scales. The speech and language development were further evaluated with meaningful use of speech scale (MUSS) and speech intelligibility rating (SIR). The postoperative speech skills were analyzed and compared between the study group and the reference group by using nonparametric statistical tests. Anatomic abnormalities of the inner ear were examined using magnetic resonance imaging (MRI) and high-resolution computed tomography of the temporal bones (HRCT).

**RESULTS:** The mean follow-up time was 14.9 years (range, 13.1-18.3 yr). Patients with DS received a multichannel implant at a mean age of 75.3 months (SD 27.9; ranging from 21 to 127 mo) and 220 non-syndromic children from reference group at a mean age of 51.4 months (SD 34.2; ranging from 9 to 167 mo). The intraoperative neural response was present in all cases. The auditory and speech performance improved in each DS child. The postoperative mean CAP and SIR scores were 4.4 (SD 0.8) and 3.2 (SD 0.6), respectively. The average of scores in MUSS and MAIS/IT-MAIS scales was 59.8% (SD 0.1) and 76.9% (SD 0.1), respectively. Gathered data indicates that children with DS implanted with CI at a younger age (<6 years of age) benefited from the CI more than children implanted later in life, similarly in a control group. There were additional anomalies of the temporal bone, external, middle, or inner ear observed in 90% of DS children, basing on MRI or HRCT.

**CONCLUSIONS:** The early cochlear implantation in children with DS is a similarly useful method in treating severe to profound sensorineural hearing loss (SNHL) as in non-syndromic patients, although the development of speech skills present differently. Due to a higher prevalence of ear and temporal bone malformations, detailed diagnostic imaging should be taken into account before the CI qualification. Better postoperative outcomes may be achieved through comprehensive care from parents/guardians and speech therapists thanks to intensive and systematic rehabilitation.

Eur J Radiol. 2020 Feb;123:108803. doi: 10.1016/j.ejrad.2019.108803. Epub 2019 Dec 26.

## **Temporal bone and intracranial abnormalities in syndromic causes of hearing loss: an updated guide.**

**D'Arco F, Youssef A, Ioannidou E, Bisdas S, Pinelli L, Caro-Dominguez P, Nash R, Siddiqui A, Talenti G.**

**PURPOSE:** To describe in detail the temporal bone and brain findings in both common and rare syndromic causes of hearing loss, with the purpose of broadening among radiologists and enhance the current understanding of distinct imaging features in paediatric patients with syndromic hearing loss.

**METHODS:** A detailed search of electronic databases has been conducted, including PubMed, Ovid Medline, Scopus, Cochrane Library, Google Scholar, National Institute for Health and Care Excellence (NICE), Embase, and PsycINFO.

**RESULTS:** Syndromic causes of hearing loss are characterised by different and sometimes specific abnormalities in the temporal bone.

**CONCLUSION:** A complete knowledge of the image findings in the temporal bones, brain, skull and other body regions is critical for the optimal assessment and management of these patients.



### [Nature and extent of hearing loss in HIV-infected children: A scoping review.](#)

[Dawood G](#), [Klop D](#), [Olivier E](#), [Elliott H](#), [Pillay M](#), [Grimmer K](#).

**INTRODUCTION:** Antiretroviral therapy (ART) has had a major impact on life expectancy from HIV as many people now live with it as a chronic disease. Chronic HIV has been associated with a range of comorbid disabilities and health conditions, one of which is hearing loss. Undiagnosed and untreated hearing loss, particularly in children, has been linked to poorer spoken language skills, with subsequent effects on academic performance.

**METHODS:** This systematic scoping review aimed to summarize the available peer-reviewed literature on hearing loss in HIV-infected children, specifically to describe its extent and nature. The review followed the framework proposed by Arksey and O'Malley. Key search terms included hearing loss (and synonyms), child (and synonyms), and HIV. Electronic databases (EBSCOhost Research Platform, PubMed, Web of Science and Scopus databases) were searched for any relevant articles published from January 1, 2000 to June 30, 2019. Reference lists of included articles were perused for additional relevant articles not already identified. Each stage of the selection process was conducted independently by two authors. The results were then collated by a third author who also resolved any discrepancies. Extracted data included sample descriptors, audiologic tests, hearing loss prevalence, hearing loss descriptors, and factors associated with hearing loss.

**RESULTS:** Seventeen articles were included; 10 from Africa, four from South America, two from North America and the remaining article from Asia. Although most of the articles reported on pure tone audiometry, the samples as well as the cut-off criteria for normal hearing were heterogeneous. Prevalence of hearing loss varied across articles (from 6% to 84%). Conductive hearing loss occurred more frequently than sensorineural or mixed hearing loss. ART use and ear infection were reported as significant in three of five articles that reported on significant associates of HIV-related hearing loss.

**CONCLUSION:** There was a modest volume of research from a limited number of countries. Heterogeneity in sampling and audiometric methods precluded a clear understanding of potential associations between chronic HIV-related hearing loss and contributing factors.

### [Cochlear Implantation in Children with Single-Sided Deafness.](#)

[Deep NL](#), [Gordon SA](#), [Shapiro WH](#), [Waltzman SB](#), [Roland JT Jr](#), [Friedmann DR](#).

**OBJECTIVE:** To describe our experience with children undergoing unilateral cochlear implantation (CI) for treatment of single-sided deafness (SSD).

**STUDY DESIGN:** Retrospective case series.

**METHODS:** A retrospective case review from a tertiary referral center involving 14 pediatric patients (<18 years) with SSD who underwent unilateral CI. Speech perception testing in quiet and noise in the CI-only and bimodal conditions with at least 1 year of device use and device usage from data logs represent the main outcome measures.

**RESULTS:** The mean age at CI was 5.0 years (median 4.4, range 1.0-11.8 years). The mean duration of deafness was 3.0 years (median 2.4, range 0.6-7.0 years). Mean follow-up was 3.4 years. Speech perception testing with a minimum of 1 year post-CI was available in eight patients. The mean word recognition scores (WRS) in the CI-only condition was 56%; a significant improvement from baseline. Testing in background noise with spatially separated speech and noise revealed that patients scored as well or better with the CI-on versus CI-off in all conditions and in no cases was interference from the CI noted. Data logs were reviewed for device usage which revealed an average use of 6.5 hr/d.

**CONCLUSION:** Cochlear implantation is a viable treatment option for pediatric SSD in this self-selected cohort. Open-set speech and improvement in background noise can be achieved. Careful patient selection and thorough counseling on expectations is paramount to achieving successful outcomes.

### [Usability of a Mobile App for Improving Literacy in Children with Hearing Impairment: A Focus Group Study.](#)

[DeForte S](#), [Sezgin E](#), [Huefner J](#), [Lucius S](#), [Luna J](#), [Satyapriya AA](#), [Malhotra P](#).

**BACKGROUND:** Children with hearing loss, even those identified early and who are using hearing aids or cochlear implants, may face challenges in developing spoken language and literacy. This can lead to academic, behavioral, and social difficulties. There are apps for healthy children to improve their spoken language and

literacy and apps that focus on sign language proficiency for children with hearing loss, but these apps are limited for children with hearing loss. We have therefore developed an app called Hear Me Read (HMR) which uses enhanced digital stories as therapy tools for speech, language, and literacy for children with hearing loss. The platform has therapist and parent/child modes that allows 1) selection of high quality, illustrated digital stories by a speech-language pathologist (SLP), parent, or child 2) modification of digital stories for a multitude of speech and language targets, and 3) assignment of stories by therapist to facilitate individualized speech and language goals. Additionally, HMR makes the caregiver a core partner in engagement through functionality whereby the caregiver can record video and audio of themselves to be played back by the child.

**OBJECTIVE:** The objective of this study was to evaluate the user experience of the HMR app through a focus group study with caregivers and their children.

**METHODS:** We recruited 16 participants (8 children with and without hearing loss and 8 caregivers) to participate in one-hour focus groups. Caregivers and children interacted with the app and discussed their experience through a semi-structured group interview. We employed thematic analysis methods and analyzed the data. We used feedback from the focus group to improve elements of the app for a larger clinical trial assessing the impact of the app on outcomes.

**RESULTS:** We identified 3 themes: default needs, specific needs and family needs. Participants found the app to be aesthetically pleasing and easy to use. Findings helped us to identify usability attributes and to amend app functionalities to best fit user needs. Caregivers and children appreciated the enhancements, such as parts of speech highlighting and video playback of caregivers reading, that were made possible by the digital format. Participants expressed that the app could be used to enhance family reading sessions and family interaction.

**CONCLUSIONS:** The findings from this focus group study are promising for the use of educational apps designed specifically for those with hearing loss who are pursuing listening and spoken language as a communication outcome. Further investigation is needed with larger sample sizes in order to understand the clinical impact on relevant language and literacy outcomes in this population.

J Pediatr. 2020 Mar;218:151-156.e2. doi: 10.1016/j.jpeds.2019.12.005. Epub 2020 Jan 14.

### [\*\*A Cross-Sectional Study of Caregiver Perceptions of Congenital Cytomegalovirus Infection: Knowledge and Attitudes about Screening.\*\*](#)

[Diener ML](#), [Shi K](#), [Park AH](#).

**OBJECTIVES:** To understand caregiver knowledge of and attitudes toward congenital cytomegalovirus (cCMV) testing in Utah.

**STUDY DESIGN:** We surveyed 365 caregivers whose children were being seen in an otolaryngology clinic at a tertiary pediatric hospital about their knowledge of and attitudes toward cCMV and cCMV screening. Descriptive statistics and cluster analysis were used to examine their responses.

**RESULTS:** The majority of caregivers were unsure how cCMV was spread, the symptoms of cCMV, and why cCMV screening of infants was important. Most caregivers did not know that cCMV screening was required by law in Utah if an infant is referred after newborn hearing screening. A majority wanted to know if their child had cCMV even if asymptomatic and were willing to pay \$20 for cCMV screening. Caregivers of children who had been tested for cCMV were significantly more likely to be strongly in favor of cCMV screening than expected by chance. Caregivers in the highly knowledgeable cluster were more likely to be strongly in favor of cCMV screening.

**CONCLUSIONS:** Caregivers frequently were unaware of cCMV and its implications. Attitudes toward cCMV screening generally were positive. Education on epidemiology and impact of cCMV may benefit both prevention of infection and attitudes toward screening.

Eur J Pediatr. 2020 May;179(5):807-812. doi: 10.1007/s00431-019-03558-7. Epub 2020 Jan 11.

### [\*\*Treatment of congenital cytomegalovirus beyond the neonatal period: an observational study.\*\*](#)

[Dorfman L](#), [Amir J](#), [Attias J](#), [Bilavsky E](#).

**ABSTRACT:** Recently, valganciclovir treatment of symptomatic congenital cytomegalovirus (cCMV) disease, commenced during the neonatal period ( $\leq 4$  weeks), was found to improve hearing and developmental outcome. However, many children (symptomatic or asymptomatic at birth) present only after 4 weeks of age. The purpose of this observational retrospective study was to describe the outcome and safety of valganciclovir therapy in infants with cCMV who started treatment  $>4$  weeks of life. Of the 91 children who started antiviral treatment  $>4$  weeks of age, 66/298 (22.2%) were symptomatic at birth; 25/217 (11.5%) were asymptomatic at birth. Treatment was initiated on average at 14 weeks of age (range 5-77 weeks) and at 53.3 weeks (range 12-156 weeks), respectively. Of the 45 affected ears in the symptomatic group, 30 (66.7%) improved and only 2 (4.4%) deteriorated, with most of the improved ears (27/30, 90%) returning to normal. In the asymptomatic group, late-onset treatment was initiated and out of the 42 deteriorated ears, 38 (90.5%) improved after at least 1 year of follow-up. Hematological adverse events, i.e., neutropenia, were noted in a minority of cases (4.4%). Conclusion: Our study demonstrates the benefits and safety aspects of treating symptomatic and asymptomatic children with

cCMV even beyond the recommended neonatal period. What is Known: • Valganciclovir treatment of symptomatic congenital cytomegalovirus (cCMV) disease, commenced during the neonatal period, is beneficial in improving hearing and developmental outcome. • However, data of treatment started beyond the neonatal period is lacking. What is New: • Our study demonstrates the benefits of treating symptomatic children with cCMV as well as asymptomatic children that develop late-onset hearing loss even beyond the recommended neonatal period. • This was true for symptomatic children who presented >4 weeks as well as to those who were asymptomatic at birth but experienced late hearing deterioration.

Eur J Hum Genet. 2020 May;28(5):587-596. doi: 10.1038/s41431-019-0553-8. Epub 2019 Dec 12.

### [Exome sequencing in infants with congenital hearing impairment: a population-based cohort study.](#)

[Downie L](#), [Halliday J](#), [Burt R](#), [Lunke S](#), [Lynch E](#), [Martyn M](#), [Poulakis Z](#), [Gaff C](#), [Sung V](#), [Wake M](#), [Hunter MF](#), [Saunders K](#), [Rose E](#), [Lewis S](#), [Jarmolowicz A](#), [Phelan D](#), [Rehm HL](#); [Melbourne Genomics Health Alliance](#), [Amor DJ](#).

**ABSTRACT:** Congenital hearing impairment (HI) is the most common sensory impairment and can be isolated or part of a syndrome. Diagnosis through newborn hearing screening and management through early intervention, hearing aids and cochlear implantation is well established in the Australian setting; however understanding the genetic basis of congenital HI has been missing. This population-derived cohort comprised infants with moderate-profound bilateral HI born in the 2016-2017 calendar years, detected through newborn hearing screening. Participants were recruited through an integrated paediatric, otolaryngology and genetics HI clinic and offered whole exome sequencing (WES) on a HiSeq4000 or NextSeq500 (Illumina) platform with a targeted average sequencing depth of 100x and chromosome microarray on the Illumina Infinium core exome-24v1.2 platform. Of those approached, 68% (106/156) consented to participate. The rate of genetic diagnosis was 56% (59/106), significantly higher than standard of care (GJB2/6 sequencing only), 21% (22/106). There were clinical implications for the 106 participants: 36% required no further screening, 9% had tailored screening initiated, 2% were offered treatment and 4% had informed care for a complex neurodevelopmental syndrome. WES in this cohort demonstrates the range of diagnoses associated with congenital HI and confirms the genetic heterogeneity of congenital HI. The high diagnostic yield and clinical implications emphasises the need for genomic sequencing to become standard of care.

Genet Med. 2020 Jan 24. doi: 10.1038/s41436-019-0745-1.

### [Exome sequencing in newborns with congenital deafness as a model for genomic newborn screening: the Baby Beyond Hearing project.](#)

[Downie L](#), [Halliday J](#), [Lewis S](#), [Lunke S](#), [Lynch E](#), [Martyn M](#), [Gaff C](#), [Jarmolowicz A](#), [Amor DJ](#).

**PURPOSE:** Genomic newborn screening raises practical and ethical issues. Evidence is required to build a framework to introduce this technology safely and effectively. We investigated the choices made by a diverse group of parents with newborns when offered tiered genomic information from exome sequencing.

**METHODS:** This population-derived cohort comprised infants with congenital deafness. Parents were offered exome sequencing and choice regarding the scope of analysis. Options were choice A, diagnostic analysis only; choice B, diagnostic analysis plus childhood-onset diseases with medical actionability; or choice C, diagnostic analysis plus childhood-onset diseases with or without medical actionability.

**RESULTS:** Of the 106 participants, 72 (68%) consented to receive additional findings with 29 (27.4%) selecting choice B and 43 (40.6%) opting for choice C. Family size, ethnicity, and age of infant at time of recruitment were the significant predictors of choice. Parents who opted to have additional findings analysis demonstrated less anxiety and decisional conflict.

**CONCLUSIONS:** These data provide evidence from a culturally diverse population that choice around additional findings is important and the age of the infant when this choice is offered impacts on their decision. We found no evidence that offering different levels of genomic information to parents of newborns has a negative psychological impact.

Int J Pediatr Otorhinolaryngol. 2020 Apr;131:109864. doi: 10.1016/j.ijporl.2020.109864. Epub 2020 Jan 7.

### [Comparison of ABR and ASSR using NB-chirp-stimuli in children with severe and profound hearing loss.](#)

[Eder K](#), [Schuster ME](#), [Polterauer D](#), [Neuling M](#), [Hoster E](#), [Hempel JM](#), [Sammelbauer S](#).

**INTRODUCTION:** Objective techniques for hearing threshold estimation in infants and children with profound or severe hearing loss play a key role in pediatric audiology to prevent speech acquisition disorders by choosing the adequate therapy. Auditory brainstem responses and auditory steady-state responses are available for frequency-dependent hearing threshold estimations and both techniques show strong correlations. However,

various systems and stimuli are available, which is one reason why comparison is challenging, and, so far, no single “gold standard” could be established for hearing threshold estimation in children suffering from profound or severe hearing loss. The aim of the study was to compare hearing threshold estimations in children with profound or severe hearing loss derived with narrow-band CE-chirps evoked auditory brainstem responses and auditory steady-state response.

**SUBJECTS and METHODS:** 71 children (121 ears) with an age from 3 month to 15 years were measured with the Interacoustics Eclipse EP25 ABR system® (Denmark) with narrow-band CE-chirps® at 500, 1000, 2000 and 4000 Hz under identical conditions.

**RESULTS:** Auditory brainstem responses and auditory steady-state responses highly correlate ( $r = 0.694$ ,  $p < 0.001$ ). Correlation coefficients differ depending on the center frequency and patient age. Generally, auditory steady-state responses show a better hearing threshold than auditory brainstem responses or a remaining hearing threshold when auditory brainstem responses could not be obtained. In approximately 15% of cases this would have affected the therapeutic strategy when only taking one technique into account.

**CONCLUSION:** Auditory brainstem responses and auditory steady-state responses should be jointly used in the diagnostic approach in children with suspected profound or severe hearing loss.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109681. doi: 10.1016/j.ijporl.2019.109681. Epub 2019 Sep 13.

### [Evaluation and therapy outcome in children with auditory neuropathy spectrum disorder \(ANSD\).](#)

[Ehrmann-Müller D](#), [Cebulla M](#), [Rak K](#), [Scheich M](#), [Back D](#), [Hagen R](#), [Shehata-Dieler W](#).

**OBJECTIVES:** The aims of the present study are to: describe diagnostic findings in patients with auditory neuropathy spectrum disorder (ANSD); and demonstrate the outcomes of different therapies like hearing aids (HAs) or cochlear implantation.

**METHODS:** 32 children were diagnosed and treated at our tertiary referral center and provided with HAs or cochlear implants (CIs). All of them underwent free-field or pure-tone audiometry. Additionally, otoacoustic emissions (OAEs), impedance measurements, auditory brainstem responses (ABRs), auditory steady-state responses (ASSR), electrocochleography, and cranial magnetic resonance imaging (cMRI) were all performed. Some patients also underwent genetic evaluation. Following suitable provision pediatric audiological tests, psychological developmental diagnostic and speech and language assessments were carried out at regular intervals in all the children.

**RESULTS:** OAEs could initially be recorded in most of the children; 17 had no ABRs. The other eight children had a poor ABR morphology. Most of the children had typical, long-oscillating cochlear microphonics (CMs) in their ABRs, which was also observed in all of those who underwent electrocochleography. Eight children were provided with a HA and 17 received a CI. The functional gain was between 32 and 65 decibel (dB) with HAs and between 32 and 50 dB with CI. A speech discrimination level between 35 and 100% was achieved during open-set monosyllabic word tests in quiet with HA or CI. With the Hochmair-Schulz-Moser (HSM) sentence test at 65 dB SPL (sound pressure level), 75% of the children with a CI achieved a speech discrimination in noise score of at least 60% at a signal to noise ratio (SNR) of 5, and four scored 80% or higher. Most of the children (72%) were full-time users of their devices. All the children with a CI used it on a regular basis.

**CONCLUSION:** Only a few case reports are available in the literature regarding the long-term outcomes of ANSD therapy. The present study reveals satisfactory outcomes with respect to hearing and speech discrimination in children with CIs or HAs. The nearly permanent use of the devices reflects a subjective benefit for the children. Provision with a suitable hearing device depends on audiological results, the speech and language development of an individual child, and any accompanying disorders. Repeated audiological evaluations, interdisciplinary diagnostics, and intensive hearing and speech therapy are essential for adequate rehabilitation of this group of children.

Blood Coagul Fibrinolysis. 2020 Apr 22. doi: 10.1097/MBC.0000000000000911.

### [Thrombosis risk of Alport syndrome patients: evaluation of cardiological, clinical, biochemical, genetic and possible causes of inherited thrombophilia and identification of a novel COL4A3 variant.](#)

[Eroz R](#), [Damar İH](#), [Kılıçaslan O](#).

**ABSTRACT:** To evaluate cases with Alport syndrome for laboratory, radiological, ophthalmological, auditory tests, cardiological and inherited thrombophilia risk. Laboratory findings, abdominal and urinary ultrasonography, ophthalmological and auditory tests and cardiological examination of 21 Alport syndrome suspicious cases

were performed. Also, collagen type IV alpha three chain (COL4A3) gene, four chain (COL4A4) gene and five chain (COL4A5) genes were sequenced by next-generation sequencing system. In addition, possible causes of inherited thrombophilia were evaluated. A novel (c.2806C>T/p.Gln936Ter) variation in COL4A3 gene was detected in three cases. Also c.221G>A/p.Arg74Gln variation in COL4A5 gene of two cases, c.4421C>T/p.Thr1474Met variation in COL4A4 gene of one case, c.665C>T/p.Pro222Leu variation in COL4A4 gene of one case and compound heterozygous c.4421C>T/(p.Thr1474Met) and c.665C>T/p.Pro222Leu variation in COL4A4 gene of one case were detected. Although 10 (47.6%) cases had microscopic hematuria, six (28.6%) cases had macroscopic hematuria, but there were not hematuria in five (23.8%) of cases. Three cases with variation carrier in COL4A genes and one case without variation carrier had vision problem. Also, one case with variation carrier in COL4A gene had hearing loss. All cases with variation carrier in COL4A genes exclude one had at least one cardiac problems. Also, all cases with variation carrier in COL4A genes had possible causes of inherited thrombophilia risk. In addition to developing risk of progressive kidney failure, sensorineural hearing loss and ocular abnormalities, Alport syndrome cases may have increasing cardiac problems and possible causes of inherited thrombophilia risk. Therefore, these cases should be regularly evaluated and followed for cardiac problems and inherited thrombophilia risk.

World J Pediatr. 2020 Jan 7. doi: 10.1007/s12519-019-00325-4.

### [Etiology of newborn hearing impairment in Guangdong province: 10-year experience with screening, diagnosis, and follow-up.](#)

[Fang BX](#), [Cen JT](#), [Yuan T](#), [Yin GD](#), [Gu J](#), [Zhang SQ](#), [Li ZC](#), [Liang YF](#), [Zeng XL](#).

**BACKGROUND:** Hearing impairment is one of the most common birth defects in children. Universal newborn hearing screenings have been performed for 19 years in Guangdong province, China. A screening/diagnosis/intervention system has gradually been put in place. Over the past 10 years, a relatively complete data management system had been established. In the present study, an etiological analysis of newborn cases that failed the initial and follow-up screenings was performed.

**METHODS:** The nature and degree of hearing impairment in newborns were confirmed by a set of procedures performed at the time of initial hearing screening, rescreening and final hearing diagnosis. Then, multiple examinations were performed to explore the associated etiology.

**RESULTS:** Over a period of 10 years, 720 children were diagnosed with newborn hearing loss. Among these children, 445 (61.81%) children had a clearly identified cause, which included genetic factor(s) (30.56%), secretory otitis media (13.30%), maternal rubella virus infection during pregnancy (5.83%), inner ear malformations (4.86%), maternal human cytomegalovirus infection during pregnancy (2.92%), malformation of the middle ear ossicular chain (2.50%) and auditory neuropathy (1.81%). In addition, 275 cases of sensorineural hearing loss of unknown etiology accounted for 38.19% of the children surveyed.

**CONCLUSIONS:** Long-term follow-up is needed to detect delayed hearing impairment and auditory development in children. The need for long-term follow-up should be taken into account when designing an intervention strategy. Furthermore, the use of the deafness gene chip should further elucidate the etiology of neonatal hearing impairment.

Am J Audiol. 2019 Dec 16;28(4):1025-1045. doi: 10.1044/2019\_AJA-19-0061. Epub 2019 Dec 12.

### [Candidacy for Amplification in Children With Hearing Loss: A Review of Guidelines and Recommendations.](#)

[Fitzpatrick EM](#), [Cologrosso E](#), [Sikora L](#).

**PURPOSE:** The 1st point in the intervention process for the majority of children is the fitting of hearing devices. The objective of this review was to compile guidelines and recommendations for candidacy criteria for children with hearing loss.

**METHOD:** Electronic databases (e.g., MEDLINE, Embase, and Cumulative Index of Nursing and Allied Health Literature) and websites were searched. Any document referring to children with hearing loss that discussed amplification guidelines or protocols was included. Documents specific to implantable devices or addressing only remote microphone systems were excluded. One reviewer screened all potentially relevant documents, and a subset was screened by a 2nd reviewer. Guidelines/recommendations referring to pediatric amplification candidacy were extracted.

**RESULTS:** A total of 40 documents were included for data extraction. Studies were categorized according to hearing loss of any degree, with separate categories for documents providing specific criteria for mild bilateral, unilateral, and auditory neuropathy spectrum disorders. Guidelines ranged from generic statements about the need for amplification to criteria based on specific audiometric thresholds. In guidelines recommending audiometric cut-points, the majority considered > 25 dB HL as a criterion for consideration for amplification. Overall, guidelines for children with mild bilateral and unilateral loss remain more ambiguous, and there was some variation across the recommendations. Guidelines for auditory neuropathy spectrum disorder stressed the need to

obtain results from behavioral audiometry before considering amplification.

**CONCLUSIONS:** Numerous organizations have established candidacy guidelines for pediatric amplification. Most guidelines specify criteria for amplification as audiometric threshold levels. There is considerable variation in the guidelines for mild bilateral and unilateral hearing loss with candidacy criteria ranging from 15 to 30 dB HL, and many guidelines recommend a case-by-case decision approach.

Int J Pediatr Otorhinolaryngol. 2020 Feb 27;133:109975. doi: 10.1016/j.ijporl.2020.109975.

### [\*\*Impact of Universal Newborn Hearing Screening on cochlear implanted children in Ireland.\*\*](#)

[Gabriel MM](#), [Geyer L](#), [McHugh C](#), [Thapa J](#), [Glynn F](#), [Walshe P](#), [Simoes-Franklin C](#), [Viani L](#).

**OBJECTIVES:** Cochlear Implant (CI) is an established treatment for severe to profound hearing loss (HL). Early diagnosis and intervention in HL are crucial in order to provide access to sound and increase the likelihood of spoken language development in pre-lingually deaf children. In April 2011, the Health Service Executive (HSE) implemented the Universal Newborn Hearing Screening (UNHS) in a phased regional basis in Ireland. This study aimed to investigate the general clinical pathway for UNHS referrals to the CI service and to evaluate the impact of earlier referrals via UNHS on functional outcomes in children.

**METHODS:** The first part of this study constituted a retrospective review of 100 children referred to the National Hearing Implant and Research Centre (NHIRC) via UNHS from November 2011 to December 2016. Implanted children referred via UNHS were categorised into three groups according to their medical status. Their clinical pathway to cochlear implantation was evaluated. Functional outcomes were investigated based on medical and developmental status, respectively. In the second part of this study, developmentally healthy implanted children referred post-UNHS were compared with medically healthy children referred pre-UNHS under the age of four, from January 2005 to June 2011. Current implant status of children, age at referral and functional outcomes were investigated.

**RESULTS:** Medically healthy children were referred to the NHIRC at an earlier age than the medically complex children (2.8 months vs 5.2 months,  $p < 0.01$ ) and the children presenting with auditory neuropathy spectrum disorder (ANSD) (2.8 months vs 5.3 months,  $p < 0.01$ ). On average they attended their first appointment and were implanted at a younger age than the ANSD group (6.1 months vs 10.1 months,  $p < 0.01$ ; 16.3 months vs 29.4 months,  $p < 0.001$ , respectively). Developmentally healthy children had significantly better functional outcomes than children with developmental delays. Children referred via UNHS were referred and implanted at a younger age than those referred pre-UNHS. The former group achieved better Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores 2 years post-implantation.

**CONCLUSION:** UNHS in Ireland is an important platform for earlier diagnosis and management of congenital HL and our results show that early intervention has a positive impact on functional outcomes in children.

J Clin Microbiol. 2020 Mar 25;58(4). pii: e01951-19. doi: 10.1128/JCM.01951-19. Print 2020 Mar 25.

### [\*\*Performance of the Alethia CMV Assay for Detection of Cytomegalovirus by Use of Neonatal Saliva Swabs.\*\*](#)

[Gantt S](#), [Goldfarb DM](#), [Park A](#), [Rawlinson W](#), [Boppa SB](#), [Lazzarotto T](#), [Mertz LM](#).

**ABSTRACT:** Congenital cytomegalovirus (cCMV) infection is a major cause of childhood hearing loss and neurodevelopmental delay. Identification of newborns with cCMV infection allows provision of beneficial interventions. However, most infants with cCMV infection have subclinical infection and go undiagnosed. Thus, expanded neonatal CMV testing is increasingly recommended. Saliva is an attractive sample type for CMV testing of newborns, because it is easier to collect than urine and more sensitive for CMV detection than dried blood spots. We evaluated the Alethia CMV assay, a rapid, easy-to-use loop-mediated isothermal amplification method for qualitative detection of CMV DNA in neonatal saliva samples. Saliva swabs were collected prospectively from newborns <21 days old and tested by the Alethia assay according to the manufacturer's instructions. Archived saliva swabs from newborns with cCMV infection were also tested retrospectively. A composite reference method (CRM; two validated PCR assays followed by bidirectional sequencing of amplicons) was performed on all samples as the reference standard comparator. Of 1,480 prospectively collected saliva swabs, 1,472 (99.5%) were negative by both the Alethia assay and CRM, 5 (0.34%) were positive by both the Alethia assay and CRM, and 3 (0.20%) were positive only by the Alethia assay. All 34 (100%) archived swabs from newborns with cCMV infection were positive by both the CRM and the Alethia assay. Overall, the Alethia assay showed 100% and 99.8% positive and negative agreement with the CRM, respectively. The Alethia CMV assay is an accurate method for identifying neonates with cCMV infection and, given its simplicity, appears suitable for CMV testing using neonatal saliva outside a reference laboratory, including remote and resource-limited settings.

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## [Screening for mitochondrial 12S rRNA C1494T mutation in 655 patients with non-syndromic hearing loss: An observational study.](#)

[Gao Z](#), [Yuan YS](#).

**ABSTRACT:** Mutations in mitochondrial DNA, especially in 12S rRNA gene, are the most important causes for hearing loss. In particular, the A1555G and C1494T mutations have been found to be associated with both aminoglycoside-induced and non-syndromic hearing loss in many families worldwide. To determine the frequency of C1494T mutation in deaf patients, in the current study, we screened this mutation in 655 patients with non-syndromic hearing loss and 300 control subjects. After PCR amplification of mitochondrial 12S rRNA gene and direct sequence analysis, we found that there were 2 patients carrying the C1494T mutation; however, this mutation was not detected in 300 healthy subjects. Further genetic counseling suggested that only 1 patient had an obvious family history of hearing impairment. Clinical evaluation showed that 3 of 10 matrilineal relatives suffered from hearing loss, with different age at onset of hearing loss. Molecular analysis revealed the presence of homoplasmic 12S rRNA C1494T and ND5 T12338C mutations, together with a set of polymorphisms belonging to human mitochondrial haplogroup F2. Interestingly, T12338C mutation resulted in the replacement of the first amino acid, a translation-initiating methionine with a threonine, shortening 2 amino acids of ND5 polypeptide. Moreover, this mutation is located in 2 nucleotides adjacent to the 3' end of the mt-tRNA<sup>Leu</sup>(CUN) gene. Therefore, this mutation may alter ND5 mRNA metabolism and the processing of RNA precursors. Thus, the combination of T12338C and C1494T mutations may contribute to deafness expression in this family. Taken together, our data suggested that the C1494T mutation was the molecular basis for hearing loss, screening for the mitochondrial DNA pathogenic mutations was recommended for early detection, prevention, and diagnosis of mitochondrial deafness.

J Perinatol. 2020 May;40(5):774-780. doi: 10.1038/s41372-020-0628-y. Epub 2020 Feb 26.

## [Treatment for hypotension in the first 24 postnatal hours and the risk of hearing loss among extremely low birth weight infants.](#)

[Gogcu S](#), [Washburn L](#), [O'Shea TM](#).

**OBJECTIVE:** To evaluate whether treated hypotension in the first 24 postnatal hours is associated with hearing loss in extremely low birth weight (ELBW) infants.

**STUDY DESIGN:** In a cohort of 735 ELBW infants, we identified 25 with sensorineural hearing loss (SNHL) at 12-24 months adjusted age. For each case, we selected three controls with normal hearing. Logistic regression models were used to adjust for confounding variables.

**RESULTS:** Sixty percent of cases and 25% of controls were treated for hypotension. After adjusting for confounding variables (gestational age, antenatal glucocorticoids, 5 min Apgar < 6, insertion of an umbilical catheter, treatment with high frequency ventilation, and major cranial ultrasound abnormality), treated hypotension was associated with an increased risk of SNHL (adjusted odds ratio: 3.6; 95% confidence interval: 1.3-9.7).

**CONCLUSIONS:** Treated hypotension in ELBW infants in the first 24 h of life is associated with an increased risk of SNHL.

Cureus. 2020 Jan 4;12(1):e6566. doi: 10.7759/cureus.6566.

## [Prevalence of Sensorineural Hearing Loss in Children with Palliated or Repaired Congenital Heart Disease.](#)

[Gopinetti L](#), [Paulpillai M](#), [Rosenquist A](#), [Van Bergen AH](#).

**BACKGROUND:** Children with congenital heart disease (CHD) are at increased risk of neurodevelopmental deficits, and the presence of sensorineural hearing loss (SNHL) may further lead to poor language skills acquisition and speech delays. Prevalence of SNHL in the general pediatric population is estimated to be 0.2% at birth to 0.35% during adolescence. Very few studies have attempted to estimate SNHL prevalence in children who have undergone congenital heart surgery.

**METHODS:** This retrospective study aimed to estimate SNHL prevalence in children who underwent congenital heart surgery in our institution and were followed up in our high-risk pediatric cardiology clinics for four years from 2009 to 2013. Data were collected on demographics, preoperative variables, surgical variables, and post-operative variables.

**RESULTS:** SNHL prevalence in asymptomatic, palliated/repared CHD patients followed in our high-risk clinics and undergoing routine surveillance was 11.6% (20 of 172 patients with hearing impairment). SNHL prevalence was not statistically higher in single-ventricle patients (17.2%) compared to biventricular patients (14.7%). Inotropic score in the first 24 hours of postoperative period ( $p=0.05$ ), lowest arterial PaO<sub>2</sub> ( $p=0.003$ ), duration of Lasix drip ( $p=0$ ), and bolus dose in days ( $p=0.03$ ) were all found to be statistically significant in the hearing-impaired group. However, using logistic regression, we identified no statistically significant predictors for hearing loss.

**CONCLUSION:** The results suggest the need for routine audiology screening of all patients with complex CHD, especially those who have undergone neonatal cardiac repair/palliation at less than one year of age, irrespective of risk factors.

Hum Genet. 2020 Apr;139(4):521-530. doi: 10.1007/s00439-020-02118-6. Epub 2020 Jan 30.

### [Concurrent hearing and genetic screening in a general newborn population.](#)

[Guo L](#), [Xiang J](#), [Sun L](#), [Yan X](#), [Yang J](#), [Wu H](#), [Guo K](#), [Peng J](#), [Xie X](#), [Yin Y](#), [Wang J](#), [Yang H](#), [Shen J](#), [Zhao L](#), [Peng Z](#).

**ABSTRACT:** Newborn hearing screening is not designed to detect delayed-onset prelingual hearing loss or aminoglycoside-antibiotic-induced ototoxicity. Cases with severe to profound hearing loss have been reported to have been missed by newborn hearing screens. The aim of this study was to evaluate the efficacy of concurrent hearing and genetic screening in the general population and demonstrate its benefits in practice. Enrolled newborns received concurrent hearing and genetic screens between September 1, 2015 and January 31, 2018. Of the 239,636 eligible infants (median age, 19 months), 548 (0.23%) had prelingual hearing loss. Genetic screening identified 14 hearing loss patients with positive genotypes and 27 patients with inconclusive genotypes who had passed the hearing screens. In addition, the genetic screen identified 0.23% (570/239,636) of the newborns and their family members as at-risk for ototoxicity, which is undetectable by hearing screens. In conclusion, genetic screening complements newborn hearing screening by improving the detection of infants at risk of hereditary hearing loss and ototoxicity, and by informing genotype-based clinical management for affected infants and their family members. Our findings suggest that the practice should be further validated in other populations and rigorous cost-effectiveness analyses are warranted.

Laryngorhinootologie. 2020 Mar 4. doi: 10.1055/a-1114-6452. [Article in German]

### [Evaluation of Universal Newborn Hearing Screening and follow-up.](#)

[Hall V](#), [Brosch S](#), [Hoffmann TK](#).

**BACKGROUND:** Universal newborn hearing screening (UNHS) was established in Germany in 2009. Even compliance was tested in early studies, there is little knowledge regarding the follow-up examination of children with suspected hearing disorder.

**METHODS:** A retrospective evaluation was performed in 570 cases of children who failed newborn hearing screening for the years between 2009-2016. Hearing deficiency was defined as having a hearing threshold  $\geq 35$ dB. Compliance with national guidelines was checked. Every child received brainstem evoked response audiometry (BERA).

**RESULTS:** Permanent hearing disorder was found in 24%, of whom about half (51%) had an inner ear hearing loss (of these in 73% bilateral). Only 27% of high risk children born in peripheral hospitals were tested immediately by the envisaged automated auditory brainstem response (AABR) method. They often presented tardy, leading to a delayed diagnosis and therapy. Children tracked by the Bavarian health office presented little earlier but had less cases who were lost to follow-up.

**DISCUSSION:** In 93% a diagnosis was made during first examination and therapy (e.g. prescription of hearing aids) initiated on average within four months age. The rate of deafness corresponded with national averages. The quality of primary screenings is crucial in revealing problems and avoiding delay in dealing with them.

J Neurooncol. 2020 Jan;146(1):147-156. doi: 10.1007/s11060-019-03356-z. Epub 2019 Nov 28.

### [Effect of sensorineural hearing loss on neurocognitive and adaptive functioning in survivors of pediatric embryonal brain tumor.](#)

[Heitzer AM](#), [Villagran AM](#), [Raghubar K](#), [Brown AL](#), [Camet ML](#), [Ris MD](#), [Hanning JH](#), [Okcu MF](#), [Paulino AC](#), [Chintagumpala M](#), [Kahalley LS](#).

**PURPOSE:** Survivors of pediatric embryonal brain tumors (BT) are at high risk for sensorineural hearing loss (SNHL) associated with neurocognitive decline. However, previous studies have not assessed the relationship between SNHL and adaptive functioning. We examined neurocognitive and adaptive functioning in patients with and without SNHL.

**METHODS:** Participants included 36 patients treated for an embryonal BT with craniospinal irradiation (CSI) and cisplatin chemotherapy who were assessed 6.7 years post-treatment on average. The impact of SNHL on neurocognitive performance and parent-rated adaptive functioning was assessed in univariate and multivariate analyses.

**RESULTS:** There were 17 cases with SNHL (mean age at evaluation = 14.4) and 19 cases with NH (mean age at evaluation = 13.8). After accounting for age at diagnosis and additional covariates in multivariable analyses, SNHL was associated with worse overall intellectual functioning ( $p=0.027$ ) and perceptual reasoning ( $p=0.016$ ) performance. There was no effect of SNHL on adaptive functioning in multivariable models. Age at diagnosis and



sex were associated with performance on neurocognitive measures.

**CONCLUSIONS:** SNHL in pediatric embryonal BT is associated with increased risk for neurocognitive deficits in conjunction with other demographic and treatment-related factors.

Cleft Palate Craniofac J. 2019 Dec 23;1055665619895635. doi: 10.1177/1055665619895635.

### [Eustachian Tube Dysfunction in Children With Unilateral Cleft Lip and Palate: Differences Between Ipsilateral and Contralateral Ears.](#)

[Hu A](#), [Shaffer AD](#), [Jabbour N](#).

**OBJECTIVE:** To evaluate Eustachian tube dysfunction in the ipsilateral and contralateral ears, in children with unilateral cleft lip and palate (UCLP).

**DESIGN:** Retrospective chart review.

**SETTING:** Tertiary care children's hospital.

**PATIENTS:** Seventy-four consecutive patients with UCLP born between 2005 and 2011 and treated at UPMC Children's Hospital of Pittsburgh Cleft-Craniofacial Center were included.

**MAIN OUTCOME MEASURES:** Conductive hearing loss, tympanogram type, number of middle ear effusions, tympanostomy tubes, and complications. Hypothesis was formulated prior to data collection.

**RESULTS:** Conductive hearing loss was nearly twice as common in the ipsilateral ear (43.2%) compared with contralateral (23.0%;  $P = .001$ , McNemar test). There were no significant differences in the frequency of each type of tympanogram between the contralateral and ipsilateral ears. The proportions of ipsilateral (90.5%) and contralateral (91.9%) ears with effusion were not significantly different. The total number of tubes received was not significantly different between the 2 ears (median of 2 bilaterally). When combined, complications (retractions, perforations, and cholesteatomas) were significantly more common in the ipsilateral ear (29.7%) compared with the contralateral ear (18.9%;  $P = .039$ , McNemar test).

**CONCLUSION:** In children with UCLP, there were significantly more instances of conductive hearing loss and complications on the cleft side compared to the noncleft side. This suggests that Eustachian tube dysfunction may indeed be more severe on the cleft side. Considering this information, clinicians may need to be especially observant of the ipsilateral ear.

Am J Audiol. 2020 Mar 5;29(1):23-34. doi: 10.1044/2019\_AJA-19-00054. Epub 2020 Jan 14.

### [Auditory Detection Thresholds and Cochlear Resistivity Differ Between Pediatric Cochlear Implant Listeners With Enlarged Vestibular Aqueduct and Those With Connexin-26 Mutations.](#)

[Jahn KN](#), [Bergan MD](#), [Arenberg JG](#).

**PURPOSE:** The goal of this study was to evaluate differences in the electrode-neuron interface as a function of hearing loss etiology in pediatric cochlear implant (CI) listeners with enlarged vestibular aqueduct (EVA) syndrome and in those with autosomal recessive connexin-26 mutations (DFNB1).

**METHOD:** Fifteen implanted ears (9 participants, 5 ears with EVA, 10 ears with DFNB1) were assessed. Single-channel auditory detection thresholds were measured using broad and spatially focused electrode configurations (steered quadrupolar; focusing coefficients = 0 and 0.9). Cochlear resistivity estimates were obtained via electrode impedances and electrical field imaging. Between-group differences were evaluated using linear mixed-effects models.

**RESULTS:** Children with EVA had significantly higher auditory detection thresholds than children with DFNB1, irrespective of electrode configuration. Between-group differences in thresholds were more pronounced on apical electrodes than on basal electrodes. In the apex, electrode impedances and electrical field imaging values were higher for children with EVA than for those with DFNB1.

**CONCLUSIONS:** The electrode-neuron interface differs between pediatric CI listeners with DFNB1 and those with EVA. It is possible that optimal clinical interventions may depend, in part, on hearing loss etiology. Future investigations with large samples should investigate individualized CI programming strategies for listeners with EVA and DFNB1.

Iran J Otorhinolaryngol. 2020 Mar;32(109):85-92. doi: 10.22038/ijorl.2019.36090.2191.

### [Prevalence of Hearing Loss among School-Age Children in the North of Iran.](#)

[Jalali MM](#), [Nezamdoost F](#), [Ramezani H](#), [Pastadast M](#).

**INTRODUCTION:** The present study aimed to investigate the audiological profiles of elementary school-age children in Rasht, Iran, and estimate the prevalence of hearing impairments in this population.

**MATERIALS AND METHODS:** In this cross-sectional descriptive-analytical study, the hearing threshold was screened using pure tone audiometry (PTA). Hearing impairment was defined as equal to or higher than 20 dB

HL. Results of the hearing thresholds were separately reported in the left or right ears and better or worse ears. Logistic regression tests were used to investigate the association between hearing loss and possible risk factors. In this study, all the analyses were conducted using SPSS software (version 21).

**RESULTS:** The present study was carried out on a total of 2019 children. Mean age of the participants was reported as  $9.66 \pm 1.66$  years. Based on low-frequency pure-tone average, the prevalence rates of hearing loss  $> 15$  dB in the right and left ears were reported as 1.94% and 1.68%, respectively. The high-frequency hearing loss  $> 15$  dB in the right and left ears was obtained at 1.14% and 1.04%, respectively. Prevalence rate of hearing loss (in all frequencies) in boys was higher than that in girls. There was a strong association between a history of otitis media and sensorineural or conductive hearing loss (adjusted odds ratio reported as 12.2 and 8.1, respectively).

**CONCLUSION:** In this study, the rate of hearing loss in the participants was approximately 2%. It was concluded that the screening of hearing loss in children is necessary for the identification and management of these children as early as possible. It is recommended to perform further trials to investigate the impact of different causes on childhood hearing impairment.

J Infect Dis. 2020 Mar 5;221(Supplement\_1):S9-S14. doi: 10.1093/infdis/jiz446.

### [Congenital Cytomegalovirus Infection.](#)

[Kabani N](#), [Ross SA](#).

**ABSTRACT:** Congenital cytomegalovirus (cCMV) infection is a leading cause of hearing loss and neurological disabilities in children, with the disease burden and disabilities due to cCMV greater than many other well recognized childhood conditions. A minority of infants with cCMV will have symptoms at birth. Infants with symptomatic cCMV are at higher risk for sequelae than those born without symptoms. The majority of infants with cCMV are asymptomatic at birth, but 10%-15% will develop hearing loss. Although clinical symptoms can help predict which infants will have sensorineural hearing loss, among asymptomatic cCMV there are currently no predictors of adverse outcome. The identification of a biomarker to identify those at highest risk of sequelae is highly desirable to target interventions to those who could potentially benefit. Because there is increasing rationale for establishing both targeted and universal screening programs for cCMV in the United States and worldwide, this is an urgent priority.

Lancet Infect Dis. 2020 Feb;20(2):220-229. doi: 10.1016/S1473-3099(19)30416-5. Epub 2019 Nov 7.

### [Congenital viral infections in England over five decades: a population-based observational study.](#)

[Kadambari S](#), [Pollard AJ](#), [Goldacre MJ](#), [Goldacre R](#).

**BACKGROUND:** Congenital viral infections cause substantial long-term morbidity but population-based data about diagnosis rates are scarce. The aim of this study was to assess the long-term trends in congenital viral infections in England and to report on how the rates of these infections might have changed with improved methods for detection, the introduction of the two-dose measles-mumps-rubella (MMR) vaccine in 1996, and the implementation of the Newborn Hearing Screening Programme (NHSP) in 2006.

**METHODS:** For this population-based, observational cohort study, we used national and regional hospitalisation data from 1968 to 2016 in England (Hospital In-Patient Enquiry, Hospital Episode Statistics, and Oxford Record Linkage Study) to calculate annual rates of hospital discharges coded with-and individuals aged younger than 1 month diagnosed with-congenital cytomegalovirus, herpes simplex virus (HSV), varicella zoster virus (VZV), and rubella. We investigated associations of congenital cytomegalovirus, HSV, and VZV with perinatal and maternal factors (sex, mother's ethnicity, mode of delivery, gestational age, birthweight, mother's age, mother's index of multiple deprivation, and number of previous pregnancies).

**FINDINGS:** In 2016, discharge rates per 100 000 infant population were 22.3 (95% CI 18.8-26.1) for congenital cytomegalovirus, 17.6 (14.6-21.1) for HSV, 32.6 (28.4-37.2) for VZV, and 0.15 (0.0-0.8) for rubella. Compared with earlier years of the study, the discharge rate in 2016 was higher for congenital cytomegalovirus, HSV, and VZV, whereas it was lower for rubella. For congenital cytomegalovirus, there was a significant step-increase between 2006 and 2007 following implementation of the NHSP (rate ratio comparing the trend line post-NHSP with that pre-NHSP 1.55 [95% CI 1.12-2.14],  $p=0.0072$ ). Congenital cytomegalovirus infection was associated with birthweight less than 1 kg, maternal age younger than 25 years, socioeconomically deprived households, caesarean section, and mothers of black ethnicity. Congenital HSV infection was associated with maternal age younger than 20 years, gestational age less than 32 weeks, and vaginal and emergency caesarean section deliveries, while VZV infection was associated with increased parity and black and south Asian ethnicities.

**INTERPRETATION:** The increase in hospital discharges coded with congenital cytomegalovirus is most likely due to the introduction of sensitive diagnostic techniques and retrospective diagnoses made in infants after implementation of the NHSP. Public health strategies to improve prevention and treatment of congenital viral infections are urgently warranted. The decrease in discharges for rubella is most likely due to the MMR vaccine.

Laryngoscope. 2020 Jan;130(1):212-216. doi: 10.1002/lary.27722. Epub 2018 Dec 8.

## [Clinical Guidelines in Pediatric Hearing Loss: Systemic Review Using the Appraisal of Guidelines for Research and Evaluation II Instrument.](#)

[Kanabur P](#), [Hubbard C](#), [Jeyakumar A](#).

**OBJECTIVES:** Despite the importance, impact, and prevalence of pediatric hearing loss (HL), there are very few published clinical practice guidelines (CPG) supporting the evaluation and management of pediatric patients with HL. Our objective was to appraise existing CPGs to ensure safe and effective practices.

**METHODS:** A literature search was conducted in PubMed, Google Scholar, EBSCO, as well as a manual Google search. Three independent assessors using the Appraisal of Guidelines for Research and Evaluation II (AGREE II) instrument evaluated CPGs related to HL in children. Standardized domain scores were calculated for each guideline.

**RESULTS:** A total of four guidelines met the inclusion criteria and were appraised. Scope and purpose achieved a high median score of 83%. Stakeholder involvement, clarity of presentation, and editorial independence achieved intermediate scores of 67%, 54%, and 50%, respectively. The areas that required most improvement and achieved low scores were rigor of development and applicability, with scores of 22% and 38%, respectively. Based on the AGREE II measures, the four guidelines had domain scores less than 60% for each domain, and without modification no guideline could be recommended.

**CONCLUSIONS:** Based on the AGREE II, the qualities of CPGs for pediatric HL have several shortcomings, and the need for a comprehensive CPG remains. Rigor of development and applicability present the greatest opportunities for improvement of these CPGs.

J Turk Ger Gynecol Assoc. 2020 Jan 13. doi: 10.4274/jtgga.galenos.2019.2019.0070.

## [Does Antenatal Magnesium Sulphate improve hearing function in premature newborns?](#)

[Kasapoğlu I](#), [Çetinkaya Demir B](#), [Atalay MA](#), [Orhan A](#), [Özkan H](#), [Çakır SC](#), [Tütüncü Toker R](#), [Kasapoğlu F](#), [Özerkan K](#).

**OBJECTIVE:** To evaluate whether antenatal magnesium sulphate (MgSO<sub>4</sub>) exposure has a neuroprotective effect against hearing impairment in premature newborns.

**MATERIALS AND METHODS:** Retrospective cohort study performed with prematurely (<37 weeks) delivered newborns at a tertiary university hospital. Newborns of 92 women who received MgSO<sub>4</sub> infusions (study group) for various indications were compared to newborns of 147 women who did not receive MgSO<sub>4</sub> infusions (control group). Every eligible premature newborn underwent hearing screening by auditory brainstem response (ABR) testing before being discharged from the hospital.

**RESULTS:** The fail rate in ABR hearing screening was 3.3% (n=3) in the study group and 10.9% (n=16) in the control group (p=0.034). The rate of concurrent use of betamethasone was higher in the study group (72.8%; n= 67) compared to control group (29.2%; n=43) (p<0.001). Other neonatal parameters such as the number of neonates who are small for gestational age and the rate of microcephaly were similar between the groups (p=0.54, p=0.48, respectively). After adjusting for co-variables including the use of betamethasone and gestational age at delivery, we did not find any statistically significant association between antenatal administration of MgSO<sub>4</sub> and fail rates in hearing screening by ABR testing (p=0.07).

**CONCLUSION:** Our results do not suggest a clear and definite benefit from antenatal MgSO<sub>4</sub> infusion in respect of hearing impairment in premature newborns.

Int J Environ Res Public Health. 2020 Apr 10;17(7). pii: E2613. doi: 10.3390/ijerph17072613.

## [What Are the Current Audiological Practices for Ototoxicity Assessment and Management in the South African Healthcare Context?](#)

[Khoza-Shangase K](#), [Masondo N](#).

**ABSTRACT:** The study was an initial exploration of the current ototoxicity assessment and management practices by audiologists in South Africa. An exploratory survey research methodology through a cross-sectional research design was adopted where audiologists were recruited from professional associations' databases in South Africa, using specific inclusion criteria. The study made use of an 18-item web-based survey guided by the Health Professions Council of South Africa (HPCSA) (2018) guidelines which were developed from reviewing international guidelines such as the American Speech-Language-Hearing Association (ASHA,1994) and the American Academy of Audiology (AAA, 2009). The study surveyed 31 audiologists from across the country. Data were analyzed through descriptive statistics. Findings implied significant gaps between knowledge and translation of this knowledge into practice. Over two thirds of the participants engage with ototoxicity monitoring and management, but the practices adopted by them do not align with international standards nor with the national HPCSA guidelines on assessment and management of patients on ototoxic medications. Most participants do not

conduct baseline assessments, and the frequency of monitoring is irregular and reduced from the recommended; thus influencing ability for early detection and intervention of ototoxicity within this context. Non-standard assessment battery is used for assessment and monitoring, raising questions about the reliability and validity of the data used to make preventive treatment decisions. Lack of collaborative work between audiologists and the rest of the clinical team involved in the treatment of patients on ototoxic medications was found to be an important contributing factor to the less than optimal ototoxicity management practices. Of factors potentially influencing adherence to guidelines, the institution of employment, specifically employment in a tuberculosis hospital, seemed to have a positive influence, possibly due to the focused nature of the audiologists' scope of practice there as well as availability of resources. The level of education appeared to have no influence. Current findings provide contextually relevant evidence on ototoxicity assessment and management within this context. They raise important implications for guidelines adherence and translating knowledge, policies and guidelines into practice, clinical assessment and management protocols followed, appropriate resource allocation per programme, as well as strategic planning for national ototoxicity assessment and management programmes in context. The findings also raise important implications for low- and middle-income countries, in terms of adopting international guidelines without considering context.

Genet Med. 2020 Mar 17. doi: 10.1038/s41436-020-0774-9.

### [Significant Mendelian genetic contribution to pediatric mild-to-moderate hearing loss and its comprehensive diagnostic approach.](#)

[Kim BJ](#), [Oh DY](#), [Han JH](#), [Oh J](#), [Kim MY](#), [Park HR](#), [Seok J](#), [Cho SD](#), [Lee SY](#), [Kim Y](#), [Carandang M](#), [Kwon IS](#), [Lee S](#), [Jang JH](#), [Choung YH](#), [Lee S](#), [Lee H](#), [Hwang SM](#), [Choi BY](#).

**PURPOSE:** Timely diagnosis and identification of etiology of pediatric mild-to-moderate sensorineural hearing loss (SNHL) are both medically and socioeconomically important. However, the exact etiologic spectrum remains uncertain. We aimed to establish a genetic etiological spectrum, including copy-number variations (CNVs) and efficient genetic testing pipeline, of this defect.

**METHODS:** A cohort of prospectively recruited pediatric patients with mild-to-moderate nonsyndromic SNHL from 2014 through 2018 (n=110) was established. Exome sequencing, multiplex ligation-dependent probe amplification (MLPA), and nested customized polymerase chain reaction (PCR) for exclusion of a pseudogene, STRCP, from a subset (n=83) of the cohort, were performed. Semen analysis was also performed to determine infertility (n=2).

**RESULTS:** Genetic etiology was confirmed in nearly two-thirds (52/83=62.7%) of subjects, with STRC-related deafness (n=29, 34.9%) being the most prevalent, followed by MPZL2-related deafness (n=9, 10.8%). This strikingly high proportion of Mendelian genetic contribution was due particularly to the frequent detection of CNVs involving STRC in one-third (27/83) of our subjects. We also questioned the association of homozygous continuous gene deletion of STRC and CATSPER2 with deafness-infertility syndrome (MIM61102).

**CONCLUSION:** Approximately two-thirds of sporadic pediatric mild-to-moderate SNHL have a clear Mendelian genetic etiology, and one-third is associated with CNVs involving STRC. Based on this, we propose a new guideline for molecular diagnosis of these children.

Clin Exp Otorhinolaryngol. 2020 Jan 14. doi: 10.21053/ceo.2019.01144.

### [A Retrospective Review of Temporal Bone Computed Tomography to Present Safe Guideline for Bone-Anchored Hearing Aids.](#)

[Kim S](#), [Cho YS](#), [Cho YS](#), [Moon IJ](#).

**OBJECTIVES:** Bone-anchored hearing device (BAHD) is contraindicated in patients younger than 5 years because their calvarial bones are not thick enough to be implanted site. However, it has not been studied in the Korean population. This study was not only to establish a safe guideline for depth of implant device in all age groups who undergo BAHD implant surgery, but also to investigate whether implantation of currently used BAHDs could be done safely in Korean children, especially those younger than 5.

**METHODS:** Two hundred eighty patients, who underwent high-resolution temporal bone computed tomography (TBCT) images between August 2010 and October 2018 were randomly enrolled in all ages. We retrospectively reviewed TBCT imaging to measure skull bone thickness at the recommended BAHD implant site.

**RESULTS:** The average skull bone thickness was 2.87 mm in patients younger than 5 years and 6.72 mm in patients older than 5 years, respectively, which conforms to the current guideline. The results indicate nearly 50% of calvarial bone thicknesses were less than 3 mm in patients under 5 years old, while 92.78% of the patients older than 5 years of age showed bone thickness greater than 4 mm. Of note, calvarial bone thickness was thicker than 3 mm in all patients who are older than 6 years.

**CONCLUSION:** This study confirms that the currently approved BAHD implantation guideline is suitable in the Korean population. For safety, we suggest taking TBCTs prior to surgery, especially in pediatric patients. Besides, noninvasive applications are recommended for patients younger than 5.

## [An Investigation of Hearing \(250-20,000 Hz\) in Children with Endocrine Diseases and Evaluation of Tinnitus and Vertigo Symptoms.](#)

[Kocuyigit M](#), [Bezgin SU](#), [Cakabay T](#), [Ortekin SG](#), [Yildiz M](#), [Ozkaya G](#), [Aydin B](#).

**INTRODUCTION:** Despite much advancement in medicine, endocrine and metabolic diseases remain an important cause of morbidity and even mortality in children.

**OBJECTIVE:** The present study was planned to investigate the evaluation of hearing that also includes high frequencies, and the presence and degree of vertigo and tinnitus symptoms in pediatric patients diagnosed with endocrine diseases such as type 1 diabetes mellitus (DM), growth hormone deficiency (GHD), obesity, idiopathic short stature, and precocious puberty

**METHODS:** The present study included a patient group of 207 children patients diagnosed with endocrine disease (95 males, 112 females; mean age 9.71 years old [range 6-16 years old]) and a control group including 55 healthy children who do not have any kind of chronic disease (26 males, 29 females; mean age 9.33 years old [range 6-16 years old]). The subjects underwent a hearing test with frequencies between 250 and 20,000 Hz. The vestibular and tinnitus symptoms were evaluated with the Pediatric Vestibular Symptom Questionnaire.

**RESULTS:** Out of 207 patients in the patient group, 5 (2.4%) had hearing loss in pure tones, 10 (4.8%) had it in high frequencies, 40 (19.3%) had tinnitus symptoms, and 18 (8.7%) had vertigo symptoms. A total of 4 out of 207 patients in the study group (1.9%), 2 out of 59 with type 1 DM patients (3.4%), 1 out of 46 with GHD (2.2%), and 1 out of 43 obesity patients (2.3%) had hearing loss, vertigo, and tinnitus symptoms.

**CONCLUSIONS:** Our results suggest that some childhood endocrine diseases can cause some changes in the inner ear, although the exact cause is unknown. Perhaps, a detailed hearing and balance examination should be a routine in a child diagnosed with an endocrine disease. We think it is necessary to work on more comprehensive patient groups and tests in the future.

## [Limitations and drawbacks of the hospital-based universal neonatal hearing screening program: First report from the Arabian Peninsula and insights.](#)

[Kolethekkat AA](#), [Al Abri R](#), [Hlaihah O](#), [Al Harasi Z](#), [Al Omrani A](#), [Sulaiman AA](#), [Al Bahlani H](#), [Al Jaradi M](#), [Mathew J](#).

**OBJECTIVES:** To assess the efficacy of the current universal neonatal hearing screening program in a tertiary medical institution in Oman, identify its limitations and drawbacks, and explore their causative factors.

**METHODS:** A retrospective review was carried out to analyse the hearing screening of 12,743 live babies born between January 2016 and December 2018. Screen coverage, drop outs, follow up rate, and age at completion of screening, diagnosis, and intervention were analysed. The results were compared with the Joint Committee on Infant Hearing (JCIH) performance quality indices. Prospective questionnaire-based telephonic interviews were then conducted with the parents or caregivers of neonates with hearing loss. Finally, the causes of loss to follow up or delays in hearing screenings, diagnosis, and/or early intervention were studied.

**RESULTS:** The true prevalence of hearing loss was 4.0 in 1000. The coverage of first-stage screening was 90% whereas the compliance with the second stage was 88.04%. 22.8% of the patients eventually obtained final diagnostic confirmation. The overall compliance with amplification was 30.2%. The completion ages of primary screening and final confirmation were 7.98 and 17.3 weeks respectively. The importance of hearing screening is well received by parents, but problems related to communication, delays in the appointment system, and inefficient follow up tracking were identified as the main limitations and drawbacks of the program.

**CONCLUSION:** The coverage of the neonatal hearing screening program had not yet reached the required goal of 95%. The performance indicators also fell below the international benchmark. There is a need to address the identified causative factors. Effective communication and well-maintained tracking systems need to be implemented.

## [The efficacy of bone-anchored hearing implant surgery in children: A systematic review.](#)

[Kruyt IJ](#), [Bakkum KHE](#), [Caspers CJI](#), [Hol MKS](#).

**OBJECTIVE:** To evaluate the efficacy of Bone-Anchored Hearing implants (BAHIs) in children and to elucidate the usage and outcomes of new surgical techniques and implants in this specific population. **DATA SOURCES:** Embase and PubMed.

**STUDY SELECTION:** We identified studies evaluating surgical outcomes of BAHIs in children. Retrieved articles were screened using predefined inclusion and exclusion criteria. Critical appraisal included directness of evidence and risk of bias. Studies that successfully passed critical appraisal were included.

**DATA EXTRACTION:** Outcome measures included patient demographics, follow-up time, surgical technique

(one-versus two-stage surgery), tissue handling technique (reduction versus preservation), type of implant used, and complications.

**DATA SYNTHESIS:** We selected 20 articles published between 2000 and 2017 for data extraction, encompassing 952 implanted BAHIs. The overall mean age at implantation was 8.6 years (range, 2-21 years). Adverse soft-tissue reactions occurred in 251 of the 952 implants (26.4%; range 0%-89% across studies). Revision surgery was performed in 16.8% (142 of the 845) of the implants. The total rate of implant loss, i.e. caused by OIF (n = 61), trauma (n = 33), recurrent infection (n = 15), elective removal due to insufficient benefit (n = 1), cosmetic reasons (n = 1), or unknown reason (n = 16), was 13.3% of the implants (127 out of 952; range 0%-40% across studies). Differences are seen in the type of implants used; wide-diameter implants seem to be superior in terms of implant survival, and similar in terms of adverse skin reactions, while one-stage surgery and soft-tissue preservation do not seem to result in higher implant loss rates or increased adverse skin reactions based upon limited amounts of literature.

**CONCLUSION:** In general, BAHIs are a safe method for hearing rehabilitation in children, although large differences between studies are observed. The outcomes of new surgical techniques and implant designs in the pediatric population seem promising, but more research is needed before definitive conclusions can be drawn.

Front Pediatr. 2020 Jan 31;8:13. doi: 10.3389/fped.2020.00013. eCollection 2020.

### [Congenital Cytomegalovirus Infection: A Narrative Review of the Issues in Screening and Management From a Panel of European Experts.](#)

[Lazzarotto T](#), [Blázquez-Gamero D](#), [Delforge ML](#), [Foulon I](#), [Luck S](#), [Modrow S](#), [Leruez-Ville M](#).

**ABSTRACT:** Maternal primary and non-primary cytomegalovirus (CMV) infection during pregnancy can result in *in utero* transmission to the developing fetus. Congenital CMV (cCMV) can result in significant morbidity, mortality or long-term sequelae, including sensorineural hearing loss, the most common sequela. As a leading cause of congenital infections worldwide, cCMV infection meets many of the criteria for screening. However, currently there are no universal programs that offer maternal or neonatal screening to identify infected mothers and infants, no vaccines to prevent infection, and no efficacious and safe therapies available for the treatment of maternal or fetal CMV infection. Data has shown that there are several maternal and neonatal screening strategies, and diagnostic methodologies, that allow the identification of those at risk of developing sequelae and adequately detect cCMV. Nevertheless, many questions remain unanswered in this field. Well-designed clinical trials to address several facets of CMV treatment (in pregnant women, CMV-infected fetuses and both symptomatic and asymptomatic neonates and children) are required. Prevention (vaccines), biology and transmission factors associated with non-primary CMV, and the cost-effectiveness of universal screening, all demand further exploration to fully realize the ultimate goal of preventing cCMV. In the meantime, prevention of primary infection during pregnancy should be championed to all by means of hygiene education.

Value Health. 2020 Feb;23(2):164-170. doi: 10.1016/j.jval.2019.07.019. Epub 2019 Oct 10.

### [Health-Related Quality of Life in Children With Low Language or Congenital Hearing Loss, as Measured by the PedsQL and Health Utility Index Mark 3.](#)

[Le HND](#), [Petersen S](#), [Mensah F](#), [Gold L](#), [Wake M](#), [Reilly S](#).

**OBJECTIVES:** To examine health-related quality of life (HRQoL) in young children with low language or congenital hearing loss and to explore

the value of assessing HRQoL by concurrently administering 2 HRQoL instruments in populations of children.

**METHODS:** Data were from 2 Australian community-based studies: Language for Learning (children with typical and low language at age 4 years, n = 1012) and the Statewide Comparison of Outcomes study (children with hearing loss, n = 108). HRQoL was measured using the parent-reported Health Utilities Index Mark 3 (HUI3) and the Pediatrics Quality of Life Inventory 4.0 (PedsQL) generic core scale. Agreement between the HRQoL instruments was assessed using intraclass correlation and Bland-Altman plots.

**RESULTS:** Children with low language and with hearing loss had lower HRQoL than children with normal language; the worst HRQoL was experienced by children with both. The lower HRQoL was mainly due to impaired school functioning (PedsQL) and speech and cognition (HUI3). Children with hearing loss also had impaired physical and social functioning (PedsQL), vision, hearing, dexterity, and ambulation (HUI3). Correlations between instruments were poor to moderate, with low agreement.

**CONCLUSIONS:** Children with low language and congenital hearing loss might benefit from interventions targeting overall health and well-being, not just their impairments. The HUI3 and PedsQL each seemed to provide unique information and thus may supplement each other in assessing HRQoL of young children, including those with low language or congenital hearing loss.

[\*\*Cytomegalovirus infection during pregnancy: state of the science.\*\*](#)

[Leruez-Ville M](#), [Foulon I](#), [Pass R](#), [Ville Y](#).

**ABSTRACT:** Cytomegalovirus is the most common congenital infection, affecting 0.5-2% of all live births and the main nongenetic cause of congenital sensorineural hearing loss and neurological damage. Congenital cytomegalovirus can follow maternal primary infection or nonprimary infection. Sensorineurological morbidity is confined to the first trimester with up to 40-50% of infected neonates developing sequelae after first-trimester primary infection. Serological testing before 14 weeks is critical to identify primary infection within 3 months around conception but is not informative in women already immune before pregnancy. In Europe and the United States, primary infection in the first trimester are mainly seen in young parous women with a previous child younger than 3 years. Congenital cytomegalovirus should be evoked on prenatal ultrasound when the fetus is small for gestation and shows echogenic bowel, effusions, or any cerebral anomaly. Although the sensitivity of routine ultrasound in predicting neonatal symptoms is around 25%, serial targeted ultrasound and magnetic resonance imaging of known infected fetuses show greater than 95% sensitivity for brain anomalies. Fetal diagnosis is done by amniocentesis from 17 weeks. Prevention consists of both parents avoiding contact with body fluids from infected individuals, especially toddlers, from before conception until 14 weeks. Candidate vaccines failed to provide more than 75% protection for >2 years in preventing cytomegalovirus infection. Medical therapies such as cytomegalovirus hyperimmune globulins aim to reduce the risk of vertical transmission but 2 randomized controlled trials have not found any benefit. Valaciclovir given from the diagnosis of primary infection up to amniocentesis decreased vertical transmission rates from 29.8% to 11.1% in the treatment group in a randomized controlled trial of 90 pregnant women. In a phase II open-label trial, oral valaciclovir (8 g/d) given to pregnant women with a mildly symptomatic fetus was associated with a higher chance of delivering an asymptomatic neonate (82%), compared with an untreated historical cohort (43%). Valganciclovir given to symptomatic neonates is likely to improve hearing and neurological symptoms, the extent of which and the duration of treatment are still debated. In conclusion, congenital cytomegalovirus infection is a public health challenge. In view of recent knowledge on diagnosis and pre- and postnatal management, health care providers should reevaluate screening programs in early pregnancy and at birth.

[\*\*Where Do We Go From Here? Some Messages to Take Forward Regarding Children With Mild Bilateral and Unilateral Hearing Loss.\*\*](#)

[Lewis DE](#).

**ABSTRACT:** This epilogue discusses messages that we can take forward from the articles in the forum. A common theme throughout the forum is the ongoing need for research. The forum begins with evidence of potential progressive hearing loss in infants with mild bilateral hearing loss, who may be missed by current newborn hearing screening protocols, and supports the need for consensus regarding early identification in this population. Consensus regarding management similarly is a continuing need. Three studies add to the growing body of evidence that children with mild bilateral or unilateral hearing loss are at risk for difficulties in speech understanding in adverse environments, as well as delays in language and cognition, and that difficulties may persist beyond early childhood. Ambivalence regarding if and when children with mild bilateral or unilateral hearing loss should be fitted with personal amplification also impacts management decisions. Two articles address current evidence and support the need for further research into factors influencing decisions regarding amplification in these populations. A third article examines new criteria to determine hearing aid candidacy in children with mild hearing loss. The final contribution in this forum discusses listening-related fatigue in children with unilateral hearing loss. The absence of research specific to this population is evidence for the need for further investigation. Ongoing research that addresses difficulties experienced by children with mild bilateral and unilateral hearing loss and potential management options can help guide us toward interventions that are specific for the needs of these children.

[\*\*A follow-up study of abnormal mutation in neonatal deafness gene screening.\*\*](#)

[Liu QM](#), [Tian Y](#), [Yu JJ](#), [He QQ](#), [Peng L](#), [Guo XQ](#), [Li DY](#), [Chen T](#).

**OBJECTIVE:** To screen, diagnose and follow up the abnormal mutation in the gene screening of neonatal deafness.

**METHODS:** A total of 24161 newborns born in Zhuhai Maternal and Child Health Hospital from February 1, 2015 to January 31, 2008 were screened for hearing and deafness genes, and audiological screening, diagnosis and 1-3 years follow-up were carried out for the newborns with positive gene screening.

**RESULTS:** There were 991 cases of deafness gene mutation (533 males and 458 females), and the rate of

abnormal mutation was 4.10%(991/24 161). Among them, 921 cases were single heterozygous mutation, 130 cases were failed in primary hearing screening, 11 cases were failed in secondary hearing screening, 8 cases were abnormal in audiological diagnosis finally. In these 8 cases, 3 were diagnosed as otitis media and passed audiological follow-up after cure, 2 cases of single ear sensorineural injury caused by high-risk factors, passed after close audiological follow-up, and the other 3 cases were closely audiological follow-up while none of them were successfully sequenced. All of them were moderate to severe sensorineural deafness, 1 case was heterozygous mutation at 3 loci of *GJB2*(c.235delC,c.408C>A,c.134G>A), 1 case was heterozygous mutation at 2 loci of *GJB2*(c.235delC, c.109G>A), and 1 case was single heterozygous mutation of *GJB2*(c.235delC). The remaining 913 cases who passed the primary screening, secondary screening or hearing diagnosis were followed up for 1 to 3 years. Three cases of multiple heterozygous mutation were found in gene screening(2 cases were *SLC26A4* 2168A>G, IVS7-2A>G, 1 case was *GJB2* c.176\_191del 16bp, c.299\_300del AT), all of them passed both primary and secondary hearing screening. In these 3 cases, the final audiological diagnosis was moderate sensorineural deafness in both ears, with no improvement in the follow-up of 1-3 years. There were 9 monogenic homozygous mutations, 7 failed in primary hearing screening, 3 failed in secondary hearing screening and also failed in audiological diagnosis and 1-3 years' audiological follow-up, all of whom were *GJB2* c.235 del C homozygous mutations, and one of whom had a definite family history of deafness. The remaining 6 cases of homozygous mutation diagnosed by primary screening, secondary screening or hearing diagnosis were *GJB2* c109G>A homozygous mutation, and passed the 1-3 years' hearing follow-up. 58 children with mtDNA mutations, including 2 with 12S rRNA 1494C>T homozygous mutation, 47 with 1555A>G homozygous mutation, and 9 with 1555A>G heterozygous mutation, all passed the primary or secondary hearing screening, and were instructed to ban ototoxic drugs for the whole life, and passed the 1-3 years' hearing follow-up.

**CONCLUSIONS:** The audiological follow-up of children with monogenic heterozygous mutations in deafness gene screening is generally normal. In case of abnormality, the influencing factors such as otitis media should be excluded at first. In case of unexplained moderate to severe sensorineural deafness, the whole-gene sequencing should be performed to find possible pathogenic factors. The children with homozygous mutation or compound heterozygous mutation in gene screening, most of whom show different degrees of hearing loss, should be followed up for a long time, and provide parents with scientific and reasonable genetic counseling according to the mutation genes and loci,. The hearing of drug-induced deafness gene carriers is normal after birth. Parents should be advised to strengthen prevention and follow-up is generally enough.

Int J Pediatr Otorhinolaryngol. 2020 Mar 12;133:109999. doi: 10.1016/j.ijporl.2020.109999.

### [Implementation of auricular malformation screenings in the newborn population.](#)

[Liu YC](#), [Kini S](#), [Barton G](#), [Pham T](#), [Marcet-Gonzalez J](#), [Novak B](#).

**BACKGROUND AND OBJECTIVE:** Research has shown that it is important to initiate ear molding early for children with auricular malformations in order to achieve the best results. Currently our institute relies on the traditional primary care physician (PCP) referral system, which does not recognize the time sensitivity of the visit in patients with auricular malformations. The purpose of the current research is to implement a new screening protocol for identifying auricular malformations in the newborn population and thus expedite the clinic visit and necessary intervention.

**METHODS:** The hearing screen technicians (HSTs) were trained to identify some of the most common auricular malformations. A picture guide of 11 types of auricular malformations were given to the HSTs to use as a reference. At the time of the newborn hearing screen, the HSTs examined the pinnae of each baby. When an auricular malformation was identified, the auricular malformation team was immediately alerted and a bedside consultation with ENT occurred.

**RESULTS:** Comparison was made of the referral rate between pre- and post-implementation of the protocol which showed an increased rate of identification (five referrals in the 12-month period pre-implementation versus eighteen referrals in the 15-month period post-implementation).

**CONCLUSION:** We successfully implemented an auricular malformation screening protocol that was linked to newborn hearing screenings. The frequency of identification has increased with the implementation of the new screening protocol and has resulted in earlier initial ENT consultations for ear molding with the goal of improving patient satisfaction and results.

Cleft Palate Craniofac J. 2020 Feb 4;1055665619899743. doi: 10.1177/1055665619899743.

### [Parental Judgement of Hearing Loss in Infants With Cleft Palate.](#)

[McAndrew L](#).

**OBJECTIVE:** To investigate whether reported parental concern is supported by hearing assessment findings in children with cleft palate. To describe this population by examining the relationship between cleft type, middle ear status, and hearing loss.

**DESIGN:** Retrospective consecutive case note review.

**SETTING:** Tertiary institutional regional cleft center.



**PATIENTS:** Consecutive cases of 194 babies born with cleft palate and referred to the specialist center from January 2009 and December 2013. Following exclusions, data from 155 infants were included for analysis.

**INTERVENTIONS:** Documented parental concern in ear, nose and throat (ENT) and speech and language therapy case notes were compared to hearing assessment findings. Findings from otoscopic examination, tympanometry, and hearing assessment were analyzed with respect to cleft type.

**RESULTS:** Parental concern is not always accurately reflected by objective assessment particularly when no concern is reported. Analysis of the cohort examined suggests that cleft type is not related to middle ear findings or hearing.

**CONCLUSIONS:** It is helpful to be aware of parental concern and clinicians should consider that parental reports may not be accurately reflected by test results. As cleft type was not found to substantially influence middle ear status or hearing it is not recommended to adapt speech and language advice offered to families according to cleft type. Follow-up studies to increase participant numbers would support a statistical analysis.

Ear Hear. 2020 Feb 12. doi: 10.1097/AUD.0000000000000829. [Epub ahead of print]  
[Prelinguistic Vocal Development in Children With Cochlear Implants: A Systematic Review.](#)  
[McDaniel J, Gifford RH.](#)

**OBJECTIVES:** This systematic review is designed to (a) describe measures used to quantify vocal development in pediatric cochlear implant (CI) users, (b) synthesize the evidence on prelinguistic vocal development in young children before and after cochlear implantation, and (c) analyze the application of the current evidence for evaluating change in vocal development before and after cochlear implantation for young children. Investigations of prelinguistic vocal development after cochlear implantation are only beginning to uncover the expected course of prelinguistic vocal development in children with CIs and what factors influence that course, which varies substantially across pediatric CI users. A deeper understanding of prelinguistic vocal development will improve professionals' abilities to determine whether a child with a CI is exhibiting sufficient progress soon after implantation and to adjust intervention as needed.

**DESIGN:** We systematically searched PubMed, ProQuest, and CINAHL databases for primary reports of children who received a CI before 5 years 0 months of age that included at least one measure of nonword, nonvegetative vocalizations. We also completed supplementary searches.

**RESULTS:** Of the 1916 identified records, 59 met inclusion criteria. The included records included 1125 total participants, which came from 36 unique samples. Records included a median of 8 participants and rarely included children with disabilities other than hearing loss. Nearly all of the records met criteria for level 3 for quality of evidence on a scale of 1 (highest) to 4 (lowest). Records utilized a wide variety of vocalization measures but often incorporated features related to canonical babbling. The limited evidence from pediatric CI candidates before implantation suggests that they are likely to exhibit deficits in canonical syllables, a critical vocal development skill, and phonetic inventory size. Following cochlear implantation, multiple studies report similar patterns of growth, but faster rates producing canonical syllables in children with CIs than peers with comparable durations of robust hearing. However, caution is warranted because these demonstrated vocal development skills still occur at older chronological ages for children with CIs than chronological age peers with typical hearing.

**CONCLUSIONS:** Despite including a relatively large number of records, the evidence in this review regarding changes in vocal development before and after cochlear implantation in young children remains limited. A deeper understanding of when prelinguistic skills are expected to develop, factors that explain deviation from that course, and the long-term impacts of variations in vocal prelinguistic development is needed. The diverse and dynamic nature of the relatively small population of pediatric CI users as well as relatively new vocal development measures present challenges for documenting and predicting vocal development in pediatric CI users before and after cochlear implantation. Synthesizing results across multiple institutions and completing rigorous studies with theoretically motivated, falsifiable research questions will address a number of challenges for understanding prelinguistic vocal development in children with CIs and its relations with other current and future skills. Clinical implications include the need to measure prelinguistic vocalizations regularly and systematically to inform intervention planning.

Int J Pediatr Otorhinolaryngol. 2020 Jan 22;132:109900. doi: 10.1016/j.ijporl.2020.109900.

[Adherence to follow-up recommendations for babies at risk for pediatric hearing loss.](#)  
[McInerney M, Scheperle R, Zeitlin W, Bodkin K, Uhl B.](#)

**OBJECTIVE:** The purpose of this retrospective study was to evaluate the families' compliance with recommendations for continued monitoring of babies with high-risk factors for hearing loss.

**METHODS:** Hearing screening and follow-up results from 604 babies were tracked across a five-year period. Bivariate analysis, including chi-square analysis, t-tests, and one-way analyses of variance were conducted to test whether various factors predicted likelihood of follow up.

**RESULTS:** Although 86% of the babies returned for the initial follow-up appointment, few completed the protocol

or were diagnosed with hearing loss (10.3%). Excluding the babies who never returned, the average age for initial assessment was near the recommended 3-month target (3.5 months). However, babies were last seen at 9.4 months on average, which is earlier than recommended. Some factors positively predicted follow-up: receipt of ototoxic medication, hyperbilirubinemia requiring transfusion, ECMO, syndromes associated with hearing loss, craniofacial anomalies, and passing the newborn hearing screening. Others were negatively predictive: NICU stay >5 days, younger maternal age, and failing the newborn screening. There was no relationship between the results of the last test and whether the families continued with monitoring. Babies with risks categorized as more likely to be associated with delayed onset hearing loss were more often late to the initial follow up, but also followed up for a longer period of time.

**CONCLUSIONS:** These results demonstrate the need to focus on the barriers unique to babies with risk factors for late onset/progressive hearing loss in addition to those barriers that generally affect loss to follow up. Tools for parental engagement are recommended.

Am J Audiol. 2019 Dec 16;28(4):823-833. doi: 10.1044/2019\_AJA-19-0047. Epub 2019 Nov 5.

## [Using Visual Supports to Facilitate Audiological Testing for Children With Autism Spectrum Disorder.](#)

[McTee HM](#), [Mood D](#), [Fredrickson T](#), [Thrasher A](#), [Bonino AY](#).

**PURPOSE:** One in 59 children is diagnosed with autism spectrum disorder (ASD). Due to overlapping symptoms between hearing loss and ASD, children who are suspected of having ASD require an audiological evaluation to determine their hearing status for the purpose of differential diagnosis. The purpose of this article is twofold: (a) to increase audiologists' knowledge of ASD by discussing the challenges associated with testing and interpreting clinical data for children with ASD or suspected ASD and (b) to provide visual supports that can be used to facilitate audiological assessment.

**METHOD:** Eight children (ages 4-12 years) were recruited as video model participants. Videos were filmed using scripts that used concise and concrete language while portraying common clinical procedures. Using the video models, corresponding visual schedules were also created.

**CONCLUSION:** Although obtaining reliable hearing data from children with ASD is challenging, incorporating visual supports may facilitate testing. Video models and visual schedules have been created and made freely available for download online under a Creative Commons License (Creative Commons-Attribution-NonCommercial-ShareAlike 4.0 International License). Incorporating visual supports during clinical testing has the potential to reduce the child's and family's stress, as well as to increase the probability of obtaining a reliable and comprehensive audiological evaluation. Future research is warranted to determine the effectiveness and feasibility of implementing these tools in audiology clinics.

Gait Posture. 2020 Mar;77:144-155. doi: 10.1016/j.gaitpost.2020.02.001. Epub 2020 Feb 3.

## [Does the practice of sports or recreational activities improve the balance and gait of children and adolescents with sensorineural hearing loss? A systematic review.](#)

[Melo RS](#), [Tavares-Netto AR](#), [Delgado A](#), [Wiesiolek CC](#), [Ferraz KM](#), [Belian RB](#).

**BACKGROUND:** Balance and gait disorders have been observed in children and adolescents with sensorineural hearing loss (SNHL), justified by vestibular dysfunctions that these children may present, due to the injury to the inner ear. Therefore, some investigations have suggested that the practice of sports or recreational activities can improve the balance and gait of this population.

**OBJECTIVE:** Assess the evidence quality from randomized or quasi-randomized controlled trials that used sports or recreational activities as an intervention to improve the balance and /or gait of children and/or adolescents with SNHL.

**METHODS:** Systematic review that surveyed articles in nine databases, published up to January 10, 2019, in any language, using the following inclusion criteria: (1) Randomized or quasi-randomized controlled trials. (2) Participants from both groups with the clinical diagnosis of SNHL, aged 6-19 years old, without physical problems, cognitive or neurological deficits, except the vestibular dysfunction. (3) Using the practice of sports or recreational activities as an intervention, to improve the balance and/or gait outcomes.

**RESULTS:** 4732 articles were identified in the searches, after the removal of the duplicates articles and the reading of the titles and their abstracts, remained 16 articles for reading in full, being 5 trials eligible for this systematic review. Of the five eligible trials, three used sports activities and two recreational activities as intervention and presented very low-quality evidence for balance and gait outcomes.

**SIGNIFICANCE:** Sports and recreational practices seem to represent promising modalities to improve the balance and gait of children and adolescents with SNHL. However, due to the methodological limitations of the trials and the low quality of the current evidence on the topic, the results of the trials should be interpreted with caution. Due to the low quality of evidence observed, we suggest that new trials be proposed on this topic, with greater methodological rigor, to provide high-quality evidence on the effectiveness of sports and recreational practices to improve the balance and gait of children and adolescents with SNHL.

## [Delayed Identification of Infants Who Are Deaf or Hard of Hearing - Minnesota, 2012-2016.](#)

[Meyer AC](#), [Marsolek M](#), [Brown N](#), [Coverstone K](#).

**ABSTRACT:** Few studies have examined factors associated with the timing of identification of hearing loss within a cohort of infants identified as deaf or hard of hearing (DHH) and what factors are associated with delayed identification. Minnesota Early Hearing Detection and Intervention (EHDI) personnel studied deidentified data from 729 infants with confirmed congenital hearing loss (i.e., hearing loss identification after not passing newborn hearing screening) born in Minnesota during 2012-2016. Differences in likelihood of delayed identification of congenital hearing loss (defined as not passing newborn hearing screening and age >3 months at the time of identification as DHH) based on multiple variables were analyzed. Overall, 222 (30.4%) infants identified as DHH had delayed identification. Multivariate regression showed that infants identified as DHH were significantly more likely to have delayed identification if they had 1) low birthweight, 2) public insurance, 3) a residence outside the metropolitan area, 4) a mother with a lower level of education, 5) a mother aged <25 years, or 6) a mother who was Hmong. Despite achievements of EHDI programs, disparities exist in timely identification of hearing loss. Using this information to develop public health initiatives that target certain populations could improve timely identification, reduce the risk for language delay, and enhance outcomes in children who are DHH.

J Perinat Med. 2020 Mar 26;48(3):234-241. doi: 10.1515/jpm-2019-0331.

## [Prenatal findings, neonatal symptoms and neurodevelopmental outcome of congenital cytomegalovirus infection in a university hospital in Montreal, Quebec.](#)

[Minsart AF](#), [Rypens F](#), [Smiljkovic M](#), [Kakkar F](#), [Renaud C](#), [Lamarre V](#), [Boucher M](#), [Boucoiran J](#).

**BACKGROUND:** Outcome of congenital cytomegalovirus (cCMV) infection in the absence of routine CMV screening and third-trimester scan in North America is scarcely documented. The aim of this study was to assess the severe outcomes related to cCMV according to the indication for screening.

**METHODS:** This was a retrospective study of 84 mother-child pairs followed for cCMV between 2003 and 2017 at CHU Sainte-Justine in Montreal, Canada. Prenatal ultrasound, neonatal symptoms, neuroimaging and severe outcomes (cerebral palsy, severe cognitive impairment, bilateral hearing loss or neonatal death) were reviewed.

**RESULTS:** Among 38 cases with abnormal prenatal ultrasound, 41.9% of live-born infants developed severe outcomes. Sixteen (42.1%) were detected in the third trimester. Among 16 cases diagnosed prenatally because of maternal history, all had normal prenatal ultrasound, and none developed severe outcomes. Among cases diagnosed postnatally because of neonatal symptoms, 25% developed severe outcomes. All infants who developed severe outcomes had moderate/severe neonatal symptoms.

**CONCLUSION:** Outcome of cCMV infection varies according to the reason for screening and timing of diagnosis. Any prenatal ultrasound anomaly might indicate a risk of severe outcome, and warrants a detailed ultrasound scan. However, late detection, or postnatal diagnosis, represented more than half of the cases, and awareness of this will help ensuring optimal management.

Am J Perinatol. 2020 Apr 10. doi: 10.1055/s-0040-1709467.

## [Auditory Brainstem Evoked Response Patterns in the Neonatal Intensive Care Unit.](#)

[Mohammed ST](#), [El-Farrash RA](#), [Taha HM](#), [Moustafa OA](#).

**OBJECTIVE:** Delayed maturation of auditory brainstem pathway in neonates admitted to the neonatal intensive care unit (NICU) may lead to misdiagnosis of children with normal peripheral hearing and inappropriate use of amplification devices. The aim of this study is to determine the pattern of auditory brain stem response in neonates admitted to the NICU for proper hearing assessment in this high-risk population.

**STUDY DESIGN:** This prospective study was conducted on 1,469 infants who were admitted to the NICU, of which 1,423 had one or more risk factors for permanent congenital hearing loss and were screened with automated auditory brain stem response (AABR). A total of 60 infants were referred for diagnostic ABR analysis after failure on AABR screening. The control group comprised 60 well-baby nursery neonates with no risk factors for PCHL.

**RESULTS:** Mean values of absolute latencies of waves III and V; interpeak latencies I-III, III-V, and I-V; amplitude of waves I, and V; and I/V amplitude ratio at 90 dBnHL measured for the right and left ears at 1 and 3 months of age show significant difference in NICU neonates compared with controls ( $p < 0.05$ ). All the diagnostic ABR measurements significantly improved at the age of 3 months ( $p < 0.001$ ) except wave I absolute latency of both groups ( $p > 0.05$ ). Significant correlations were found between ABR readings at the age of 1 and 3 months and the gestational age of the NICU neonates ( $p < 0.05$ ).

**CONCLUSION:** Diagnostic ABR findings in NICU neonates suggested delayed maturation of the auditory brainstem pathway with a great impact of gestational age on this maturation. Auditory maturational changes were observed at 3 months of age of patient and control groups.

[Natural History of Congenital Cytomegalovirus Infection in Highly Seropositive Populations.](#)  
[Mussi-Pinhata MM, Yamamoto AY.](#)

**ABSTRACT:** Maternal preconceptional cytomegalovirus (CMV) immunity does not protect the fetus from acquiring congenital CMV infection (cCMV). Nonprimary infections due to recurrence of latent infections or reinfection with new virus strains during pregnancy can result in fetal infection. Because the prevalence of cCMV increases with increasing maternal CMV seroprevalence, the vast majority of the cases of cCMV throughout the world follow nonprimary maternal infections and is more common in individuals of lower socioeconomic background. Horizontal exposures to persons shedding virus in bodily secretions (young children, sexual activity, household crowding, low income) probably increase the risk of acquisition of an exogenous nonprimary CMV infection and fetal transmission. In addition, more frequent acquisition of new antibody reactivities in transmitter mothers suggest that maternal reinfection by new viral strains could be a major source of congenital infection in such populations. However, the exact frequency of CMV nonprimary infection in seroimmune women during pregnancy and the rate of intrauterine transmission in these women are yet to be defined. Usually, the birth prevalence of cCMV is high ( $\geq 7:1000$ ) in highly seropositive populations. There is increasing evidence that the frequency and severity of the clinical and laboratory abnormalities in infants with congenital CMV infection born to mothers with nonprimary CMV infection are similar to infants born after a primary maternal infection. This is particularly true for sensorineural hearing loss, which contributes to one third of all early-onset hearing loss in seropositive populations. This brief overview will discuss the need for more research to better clarify the natural history of cCMV in highly seropositive populations, which, in almost all populations, remains incompletely defined.

[Comparison of the Pediatric Cochlear Implantation Using Round Window and Cochleostomy.](#)  
[Naderpour M, Aminzadeh Z, Jabbari Moghaddam Y, Pourshiri B, Ariafar A, Akhondi A.](#)

**INTRODUCTION:** Cochlear implantation (CI) is now regarded as a standard treatment for children with severe to profound sensor neural hearing loss. This study aimed to compare the efficacy of the round window approach (RWA) and standard cochleostomy approach (SCA) in the preservation of residual hearing after CI in pediatric patients.

**MATERIALS AND METHODS:** This double-blind randomized controlled trial was conducted on 97 pediatric patients receiving CI with 12-month follow-up. The study population was divided into two groups according to the surgical approaches they received, including RWA and SCA. Consequently, the patients were evaluated based on the Categories of Auditory Performance scale (CAP) and Speech Intelligibility Rating (SIR) test 45-60 days and 3, 6, 9, and 12 months post-surgery.

**RESULTS:** The CAP and SIR mean scores increased in both groups during the 12-month follow-up. This upward trend was significant in both groups ( $P < 0.001$ ). There was no significant difference between the two treatment groups in any of the follow-up stages regarding the CAP mean score. The mean SIR score ( $P = 1.14 \pm 0.40$ ) was significantly higher in the RWA group 3 ( $P = 0.001$ ), 6 ( $P = 0.008$ ), and 9 ( $P = 0.006$ ) months after the surgery. However, there was no significant difference between the RWA and SCA groups, regarding 1-year SIR ( $P = 0.258$ ).

**CONCLUSION:** The CI with either RWA or SCA could improve hearing and speech performance in pediatric patients. Although mid-term speech intelligibility was better for RWA, there was no significant difference in the 1-year outcome between these two methods.

[Characterization of a universal screening approach for congenital CMV infection based on a highly-sensitive, quantitative, multiplex real-time PCR assay.](#)

[Nagel A, Dimitrakopoulou E, Teig N, Kern P, Lücke T, Michna D, Korn K, Steininger P, Shahada K, Neumann K, Überla K.](#)

**ABSTRACT:** The majority of congenital cytomegalovirus (cCMV) infections are asymptomatic at birth and therefore not diagnosed. Approximately 10-15% of these infants develop late-onset hearing loss and other developmental disorders. Implementation of a universal screening approach at birth may allow early initiation of symptomatic interventions due to a closer follow-up of infants at risk and offers the opportunity to consider treatment of late-onset disease. Real-time PCR assays for the detection of CMV DNA in buccal swab samples demonstrated feasibility and good clinical sensitivity in comparison to a rapid culture screening assay. Because most cCMV infections remain asymptomatic, a universal screening assay that stratifies CMV infected infants according to low and high risk of late-onset cCMV disease could limit the parental anxiety and reduce follow-up costs. We therefore developed and characterized a screening algorithm based on a highly-sensitive quantitative real-time PCR assay that is compatible with centralized testing of samples from universal screening and allows to

determine CMV DNA load of saliva samples either as International Units (IU)/ml saliva or IU/105 cell equivalents. 18 of 34 saliva samples of newborns that tested positively by the screening algorithm were confirmed by detection of CMV DNA in blood and/or urine samples obtained during the first weeks of life. All screening samples that could not be confirmed had viral loads of  $<2.3 \times 10^5$  IU/ml saliva (median:  $6.8 \times 10^3$ ) or  $1.3 \times 10^5$  IU/105 cell equivalents (median:  $4.0 \times 10^2$ ). The viral load of screening samples with confirmed cCMV infection ranged from  $7.5 \times 10^2$  to  $8.2 \times 10^9$  IU/ml saliva (median:  $9.3 \times 10^7$ ) or  $1.5 \times 10^2$  to  $5.6 \times 10^{10}$  IU/105 cell equivalents (median:  $3.5 \times 10^6$ ). Clinical follow-up of these newborns with confirmed cCMV infection should reveal whether the risk of late-onset cCMV disease correlates with CMV DNA load in early life saliva samples and whether a cut-off can be defined identifying cCMV infected infants with or without risk for late-onset cCMV disease.

Otolaryngol Head Neck Surg. 2020 Mar;162(3):319-321. doi: 10.1177/0194599819900492. Epub 2020 Jan 21.

[\*\*Letters to the Deaf: Present-Day Relevance of History's Earliest Social Analysis of Deafness.\*\*](#)  
[Naples J, Valdez TA.](#)

**ABSTRACT:** Harriet Martineau was a 19th-century sociologist who had a progressive form of deafness. Her 1834 essay, *Letters to the Deaf*, was the earliest historical document depicting the social challenges of hearing loss. Martineau details complex situations that hard-of-hearing people experienced in the 19th century such as social isolation due to frustrations with communication, physician shortcomings, limited music appreciation, and the stigma of hearing amplification devices. Her descriptions of these experiences are commonly faced by hard-of-hearing people in present-day society. Advancements in technology and recognition of the negative social impact of hearing loss have improved the social experience for the hard of hearing; however, social challenges remain relevant. In this article, we review *Letters to the Deaf* and note the ways in which this essay provides a dual perspective regarding how much we have advanced as a society and how much we still have to overcome in addressing the social challenges of hearing loss.

Acta Otorrinolaringol Esp. 2020 Jan - Feb;71(1):45-55. doi: 10.1016/j.otorri.2018.09.004. Epub 2018 Dec 19.

[\*\*Early diagnosis and treatment of unilateral or asymmetrical hearing loss in children: CODEPEH recommendations.\*\*](#)

[Núñez-Batalla F, Jáudenes-Casaubón C, Sequí-Canet JM, Vivanco-Allende A, Zubicaray-Ugarteche J.](#)

**ABSTRACT:** The aim of this document is to improve the management and the treatment of unilateral or asymmetrical hearing loss in children. One in one thousand newborn infants has unilateral hearing loss and this prevalence increases with age, due to cases of acquired and delayed-onset hearing loss. Although the impact on the development and learning processes of children of these kinds of hearing loss have usually been minimized, if they are not treated they will impact on language and speech development, as well as overall development, affecting the quality of life of the child and his/her family. The outcomes of the review are expressed as recommendations aimed at clinical diagnosis and therapeutic improvement for unilateral or asymmetrical hearing loss.

Acta Med Port. 2019 Dec 2;32(12):767-775. doi: 10.20344/amp.11880. Epub 2019 Dec 2. [Article in Portuguese]

[\*\*Congenital or Early Acquired Deafness: An Overview of the Portuguese Situation, from Diagnosis to Follow-Up.\*\*](#)

[Oliveira C, Machado M, Zenha R, Azevedo L, Monteiro L, Bicho A.](#)

**INTRODUCTION:** Congenital deafness or early acquired deafness affects 1 to 3 out of 1000 newborns without risk factors and 20 to 40 out of 1000 newborns with risk factors. The universal newborn hearing screening enables its early identification. Children with congenital deafness/early acquired deafness have a higher prevalence of other conditions, especially ophthalmologic and neurodevelopmental ones, and at least 30% to 40% have at least one associated comorbidity.

**MATERIAL AND METHODS:** We carried out a cross-sectional, multicenter study in which 83% (n = 30) of the hospitals/maternity hospitals of the National Health Service participated.

**RESULTS:** All surveyed hospitals/maternity hospitals routinely performed universal newborn hearing screening to all newborns before discharge; 63% referred children with risk factors for hearing loss to Otorhinolaryngology. All children with congenital deafness/early acquired deafness are referred to: Pediatrics in 23% hospitals/maternity hospitals. In 23 hospitals/maternity hospitals, all children with congenital deafness/early acquired deafness are referred to: Speech Therapy in 44% hospitals/ maternity hospitals; Ophthalmology in 17% hospitals/maternity hospitals; National System of Early Intervention in Childhood in 30% hospitals/maternity hospitals; 22% of hospitals/maternity hospitals refer all children with congenital deafness/early acquired deafness, with no identified cause, to Clinical Genetics clinics. The number of diagnoses of deafness in the years 2014 and 2015 was 2.5 and 1.5 per 1000 newborns, respectively, in 15 hospitals/maternity hospitals.

**DISCUSSION:** Awareness of universal newborn hearing screening seems to be widely spread in the National Health Service. The number of children with SC / SPA, as well as the percentage of different types of deafness

diagnosed, were identical to those found in other studies and shows its importance. The assessment / follow-up of these children by specialties other than the otolaryngology was heterogeneous in different health entities and revealed that not all children with risk factors for deafness follow up advised by existing standards.

**CONCLUSION:** Results show that Portugal made an important path in the screening and follow-up of children with SC / SPA. It is important, with the ultimate aim of continually improving the care of these children, to reflect on the involvement of specialties other than otolaryngology, such as the National Early Childhood Intervention System in the follow-up of these children.

Indian J Otolaryngol Head Neck Surg. 2020 Mar;72(1):30-35. doi: 10.1007/s12070-019-01723-w. Epub 2019 Jul 31.

### [A Prospective Study on Temporal Bone Involvement in Polytrauma Patients and the Effect of Early Diagnosis on Hearing Loss.](#)

[Padmakumar V](#), [Ramesh Kumar E](#), [Ramakrishnan VR](#).

**ABSTRACT:** As polytrauma cases are on the rise, a large number of patients presents with temporal bone fractures, which can result in various types of injuries varying from trivial to more serious injuries. Early diagnosis and appropriate management is required in case of serious injuries for a better outcome. The aim of my study is to study the incidence, the different injuries occurring and the effect of early diagnosis on hearing loss. Patients coming to our emergency department with polytrauma are studied and clinically evaluated for any temporal bone injuries. Based on the type of injuries audiological and radiological studies are done. And if required, biochemical tests like CSF analysis will be done. Also hearing assessment will be done as early as possible and appropriate treatment required will be started. The outcome is then assessed and followed up on a regular basis. In our study there were 90 patients with temporal bone fracture out of the 2748 polytrauma cases. The incidence was calculated to be 32 per 1000 cases. 69 patients (76.7%) had longitudinal fracture of temporal bone; 13 patients (14.4%) had transverse fracture; 2 patients (2.2%) had oblique fractures and 6 patients (6.6%) had comminuted fractures. Hearing loss was found to be the most common injury seen in 56 patients (62.2%). Of which 30 (53.5%) had conductive hearing loss (CHL); 9 (16%) had sensorineural hearing loss (SNHL); 17 had mixed hearing loss (MHL). 27 (90%) out of 30 patients with CHL showed improvement in hearing. Out of the 26 patients with SNHL and MHL, 22 patients (84.61%) showed improvement. 5 out of 6 with immediate onset facial palsy and 6 out of 8 with late onset facial palsy showed improvement. The hearing outcome in our study was found to be much better than the previous year which shows that the difference might be due to the early diagnosis and management. In our study hearing improvement was noted in most patients with hearing loss when compared to the previous year, which may have been due to the detection of the injuries at the earliest and managing the same with appropriate treatment modalities.

Mol Genet Genomic Med. 2020 Feb 17:e1171. doi: 10.1002/mgg3.1171.

### [Analyses of del\(GJB6-D13S1830\) and del\(GJB6-D13S1834\) deletions in a large cohort with hearing loss: Caveats to interpretation of molecular test results in multiplex families.](#)

[Pandya A](#)<sup>1</sup>, [O'Brien A](#)<sup>1</sup>, [Kovasala M](#)<sup>1</sup>, [Bademci G](#)<sup>2</sup>, [Tekin M](#)<sup>2</sup>, [Arnos KS](#)<sup>3</sup>.

**BACKGROUND:** Mutations involving the closely linked GJB2 and GJB6 at the DFNB1 locus are a common genetic cause of profound congenital hearing loss in many populations. In some deaf GJB2 heterozygotes, a 309 kb deletion involving the GJB6 has been found to be the cause for hearing loss when inherited in trans to a GJB2 mutation.

**METHODS:** We screened 2,376 probands from a National DNA Repository of deaf individuals.

**RESULTS:** Fifty-two of 318 heterozygous probands with pathogenic GJB2 sequence variants had a GJB6 deletion. Additionally, eight probands had an isolated heterozygous GJB6 deletion that did not explain their hearing loss. In two deaf subjects, including one proband, a homozygous GJB6 deletion was the cause for their hearing loss, a rare occurrence not reported to date.

**CONCLUSION:** This study represents the largest US cohort of deaf individuals harboring GJB2 and GJB6 variants, including unique subsets of families with deaf parents. Testing additional members to clarify the phase of GJB2/GJB6 variants in multiplex families was crucial in interpreting clinical significance of the variants in the proband. It highlights the importance of determining the phase of GJB2/GJB6 variants when interpreting molecular test results especially in multiplex families with assortative mating.

BMC Infect Dis. 2020 Mar 12;20(1):217. doi: 10.1186/s12879-020-4941-z.

### [Diagnosing congenital Cytomegalovirus infection: don't get rid of dried blood spots.](#)

[Pellegrielli L](#), [Alberti L](#), [Pariani E](#), [Barbi M](#), [Binda S](#).

**BACKGROUND:** Congenital Cytomegalovirus (cCMV) is a serious global public health issue that can cause irreversible fetal and neonatal congenital defects in symptomatic or asymptomatic newborns at birth. In absence

of universal cCMV screening, the retrospective diagnosis of cCMV infection in children is only possible by examining Dried Blood Spot (DBS) samples routinely collected at birth and stored for different time spans depending on the newborn screening regulations in force in different countries. In this article, we summarize the arguments in favor of long-term DBS sample storage for detecting cCMV infection.

**MAIN TEXT:** CMV infection is the most common cause of congenital infection resulting in severe defects and anomalies that can be apparent at birth or develop in early childhood. Sensorineural hearing loss is the most frequent consequence of cCMV infection and may have a late onset and progress in the first years of life. The virological diagnosis of cCMV is essential for clinical research and public health practices. In fact, in order to assess the natural history of CMV infection and distinguish between congenital or acquired infection, children should be diagnosed early by analyzing biological samples collected in the first weeks of life (3 weeks by using viral culture and 2 weeks by molecular assays), which, unfortunately, are not always available for asymptomatic or mildly symptomatic children. It now seems possible to overcome this problem since the CMV-DNA present in the blood of congenitally infected newborns can be easily retrieved from the DBS samples on the Guthrie cards routinely collected and stored within 3 days from birth in the neonatal screening program for genetic and congenital diseases. Early collection and long-term storage are inexpensive methods for long-term bio-banking and are the key points of DBS testing for the detection of cCMV.

**CONCLUSION:** DBS sampling is a reliable and inexpensive method for long-term bio-banking, which enables to diagnose known infectious diseases - including cCMV - as well as diseases not yet recognized, therefore their storage sites and long-term storage conditions and durations should be the subject of political decision-making.

Laryngoscope. 2020 Jan 27. doi: 10.1002/lary.28536.

### [Genetic Testing for Congenital Bilateral Hearing Loss in the Context of Targeted Cytomegalovirus Screening.](#)

[Peterson J](#), [Nishimura C](#), [Smith RJH](#).

**OBJECTIVES/HYPOTHESIS:** To determine the prevalence of children with genetic hearing loss who are cytomegalovirus (CMV) positive at birth and the relative proportion of genetic and CMV etiology among children with congenital bilateral hearing loss.

**STUDY DESIGN:** Database review.

**METHODS:** We performed a review of clinical test results for patients undergoing comprehensive genetic testing for all known hearing loss-associated genes from January 2012 to January 2019. This population was reviewed for reported CMV status and genetic causes of congenital bilateral hearing loss.

**RESULTS:** In the OtoSCOPE database, 61/4,282 patients were found to have a documented CMV status, and 661/4282 had documented bilateral congenital hearing loss. Two patients were identified who had both a positive CMV result and a genetic cause for their hearing loss. Forty-eight percent of patients with bilateral congenital hearing loss (320/661) were found to have a genetic etiology. In 62% (198/320), the hearing loss was associated with pathogenic variants in GJB2, STRC, SLC26A4 or an Usher syndrome-associated gene.

**CONCLUSIONS:** We estimate that ~2% of CMV-positive newborns with hearing loss have a known genetic variant as a cause. The subcohort of CMV-positive newborns with symmetric mild-to-moderate bilateral hearing loss will have at least a 7% chance of having pathogenic gene variants associated with hearing loss. In a CMV-positive neonate who failed their newborn hearing screen bilaterally, genetic screening needs to be considered for accurate diagnosis and possible deferment of antiviral treatment.

J Pediatr Hematol Oncol. 2020 Jan;42(1):e25-e31. doi: 10.1097/MPH.0000000000001637.

### [Platinum-drugs Ototoxicity in Pediatric Patients With Brain Tumors: A 10-Year Review.](#)

[Rabiço-Costa D](#), [Gil-da-Costa MJ](#), [Barbosa JP](#), [Bom-Successo M](#), [Spratley J](#).

**PURPOSE:** Platinum-derived chemotherapy is one of the cornerstones in the treatment of central nervous system tumors in children. We aimed to assess the incidence of hearing loss in children after the exposure to platinum drugs.

**MATERIAL AND METHODS:** Retrospective study of prospectively collected data on children consecutively diagnosed with brain tumors and treated with platinum derivatives at a tertiary referral hospital between January 2006 and December 2015. We analyzed multiples variables, such as: age at diagnosis, tumor location, hydrocephalus, platinum drug type, radiotherapy, and follow-up time. The final sample size was 51 patients.

**RESULTS:** The median age at diagnosis was 6 years. The median overall follow-up time was 75 months. The incidence of ototoxicity was 23.5%. Rates of hearing loss with carboplatinum were lower than with cisplatinum. A statistically significant association occurred between the presence of hydrocephalus, radiotherapy exposure, infratentorial tumor location, and ototoxicity after treatment with platinum derivatives.

**CONCLUSIONS:** Childhood central nervous system tumors nowadays exhibit improved cure and survival rates. However, the ototoxicity resulting from the chemotherapy treatment may accompany patients for the rest of their lives. This study reveals that this occurrence is not negligible, and the association of radiotherapy and the presence of hydrocephalus can be potentiating factors.

### [Identification of TMC1 as a relatively common cause for nonsyndromic hearing loss in the Saudi population.](#)

[Ramzan K](#), [Al-Owain M](#), [Al-Numair NS](#), [Afzal S](#), [Al-Ageel S](#), [Al-Amer S](#), [Al-Baik L](#), [Al-Otaibi GF](#), [Hashem A](#), [Al-Mashharawi E](#), [Basit S](#), [Al-Mazroea AH](#), [Softah A](#), [Sogaty S](#), [Imtiaz F](#).

**ABSTRACT:** Hearing loss (HL) is the most common sensory disorder worldwide and genetic factors contribute to approximately half of congenital HL cases. HL is subject to extensive genetic heterogeneity, rendering molecular diagnosis difficult. Mutations of the transmembrane channel-like 1 (TMC1) gene cause hearing defects in humans and mice. The precise function of TMC1 protein in the inner ear is unknown, although it is predicted to be involved in functional maturation of cochlear hair cells. TMC1 mutations result in autosomal recessive (DFNB7/11) and sometimes dominant (DFNA36) nonsyndromic HL. Mutations in TMC1 are responsible for a significant portion of HL, particularly in consanguineous populations. To evaluate the importance of TMC1 mutations in the Saudi population, we used a combination of autozygome-guided candidate gene mutation analysis and targeted next generation sequencing in 366 families with HL previously shown to lack mutations in GJB2. We identified 12 families that carried five causative TMC1 mutations; including three novel (c.362+3A>G; c.758C>T [p.Ser253Phe]; c.1396\_1398delACC [p.Asn466del]) and two reported mutations (c.100C>T [p.Arg34Ter]; c.1714G>A [p.Asp572Asn]). Each of the identified recessive mutation was classified as severe, by both age of onset and severity of HL. Similarly, consistent with the previously reported dominant variant p.Asp572Asn, the HL phenotype was progressive. Eight families in our cohort were found to share the pathogenic p.Arg34Ter mutation and linkage disequilibrium was observed between p.Arg34Ter and SNPs investigated. Our results indicate that TMC1 mutations account for about 3.3% (12/366) of Saudi HL cases and that the recurrent TMC1 mutation p.Arg34Ter is likely to be a founder mutation.

### [Structural neuroimaging of the altered brain stemming from pediatric and adolescent hearing loss-Scientific and clinical challenges.](#)

[Ratnanather JT](#).

**ABSTRACT:** There has been a spurt in structural neuroimaging studies of the effect of hearing loss on the brain. Specifically, magnetic resonance imaging (MRI) and diffusion tensor imaging (DTI) technologies provide an opportunity to quantify changes in gray and white matter structures at the macroscopic scale. To date, there have been 32 MRI and 23 DTI studies that have analyzed structural differences accruing from pre- or perilingual pediatric hearing loss with congenital or early onset etiology and postlingual hearing loss in pre-to-late adolescence. Additionally, there have been 15 prospective clinical structural neuroimaging studies of children and adolescents being evaluated for cochlear implants. The results of the 70 studies are summarized in two figures and three tables. Plastic changes in the brain are seen to be multifocal rather than diffuse, that is, differences are consistent across regions implicated in the hearing, speech and language networks regardless of modes of communication and amplification. Structures in that play an important role in cognition are affected to a lesser extent. A limitation of these studies is the emphasis on volumetric measures and on homogeneous groups of subjects with hearing loss. It is suggested that additional measures of morphometry and connectivity could contribute to a greater understanding of the effect of hearing loss on the brain. Then an interpretation of the observed macroscopic structural differences is given. This is followed by discussion of how structural imaging can be combined with functional imaging to provide biomarkers for longitudinal tracking of amplification.

### [Clinical Diagnostic Testing for Human Cytomegalovirus Infections.](#)

[Razonable RR](#), [Inoue N](#), [Pinninti SG](#), [Boppana SB](#), [Lazzarotto T](#), [Gabielli L](#), [Simonazzi G](#), [Pellett PE](#), [Schmid DS](#).

**ABSTRACT:** Human cytomegalovirus (HCMV) infections are among the most common complications arising in transplant patients, elevating the risk of various complications including loss of graft and death. HCMV infections are also responsible for more congenital infections worldwide than any other agent. Congenital HCMV (cCMV) infections are the leading nongenetic cause of sensorineural hearing loss and a source of significant neurological disabilities in children. While there is overlap in the clinical and laboratory approaches to diagnosis of HCMV infections in these settings, the management, follow-up, treatment, and diagnostic strategies differ considerably. As yet, no country has implemented a universal screening program for cCMV. Here, we summarize the issues, limitations, and application of diagnostic strategies for transplant recipients and congenital infection, including examples of screening programs for congenital HCMV that have been implemented at several centers in Japan, Italy, and the United States.



**Impact of cochlear abnormalities on hearing outcomes for children with cochlear implants.**

Ronner E, Basonbul R, Bhakta R, Mankarious L, Lee DJ, Cohen MS.

**OBJECTIVE:** Evaluate the impact of cochlear anomalies on hearing outcomes for pediatric patients with cochlear implants.

**STUDY DESIGN:** Retrospective chart review.

**SETTING:** Tertiary care center.

**SUBJECTS AND METHODS:** Charts were retrospectively reviewed for cases where pediatric cochlear implant surgery was performed between 2002 and 2018 at a single, tertiary care institution. Patients were divided into groups based on the presence or absence of radiological cochlear abnormalities, which were further classified as low or high risk anomalies. Hearing outcomes were evaluated by measuring pure tone averages and word recognition scores preoperatively, 3 and 12 months postoperatively, in addition to the most recent test results.

**RESULTS:** There were 154 ears implanted in our cohort of 100 patients. 107 ears had normal cochlear anatomy, 31 had low risk, and 16 had high risk abnormalities. The most common modality of preoperative imaging was CT scan. Postoperative mean pure tone average (PTA) was significantly higher in patients with inner ear anomalies compared to those with normal anatomy. No significant difference in PTA was noted between low versus high risk patients. <50% of patients had word recognition scores available within the first year following surgery.

**CONCLUSION:** Abnormalities of the inner ear significantly influenced hearing outcomes over time following cochlear implant surgery when compared to pediatric patients with normal anatomy. Obtaining hearing testing can be difficult in very young children and therefore future studies are warranted to further investigate the impact that cochlear abnormalities may have on hearing outcomes following cochlear implant surgery.

**Outcomes of regional-based newborn hearing screening for 35,461 newborns for 5 years in Akita, Japan.**

Sato T, Nakazawa M, Takahashi S, Mizuno T, Ishikawa K, Yamada T.

**OBJECTIVES:** Newborn hearing screening (NHS) has been actively performed since 2001 in Akita, Japan. The NHS coverage rate has increased yearly, and performance has been consistently >90% since 2012. The purpose of this study was to summarize NHS outcomes in the Akita prefecture of Japan and to obtain new insights for from our summarized data for the future.

**METHODS:** A total of 35,461 newborns in hospitals and clinics where hearing screening was performed in Akita from 2012 to 2016 were included. The outcome data of NHS were collected for analysis.

**RESULTS:** The overall screening coverage rate for hearing loss was 94.7%. Of the screened infants, 0.53% received a referral on the 2-stage automated auditory brainstem response (ABR), and 80.4% of referred infants had a check-up at the hospital to receive a diagnostic hearing examination. Finally, the prevalence of bilateral congenital hearing loss was 0.14%, that of bilateral moderate to profound hearing loss was 0.12%, and that of unilateral congenital hearing loss was 0.10%. Furthermore, the average consultation period in infants with risk factors was significantly later than that in infants without risk factors ( $p = 0.0015$ ). Follow-up for infants diagnosed with normal hearing after diagnostic hearing examination revealed that 4.7% suffered bilateral moderate to profound hearing loss later. This percentage is significantly higher than that of the general group ( $p < 0.001$ ).

**CONCLUSION:** The prevalence of bilateral congenital hearing loss was 0.14% in Akita and 0.12% of infants were diagnosed with bilateral moderate to severe hearing loss. Medical personnel should be enlightened regarding the importance of performing hearing diagnostic examinations until 3 months of age. Even if infants were diagnosed with normal hearing after a diagnostic examination, we strongly suggest continuing follow-up until they are able to perform pure tone audiometry with accuracy.

**Guideline: Auditory processing and perception disorders: Proposal for treatment and management of APD : S1 guideline of the German Society of Phoniatics and Pediatric Audiology.**

Schönweiler R, Kiese-Himmel C, Plotz K, Nickisch A, Am Zehnhoff-Dinnesen A.

**ABSTRACT:** Despite normal hearing thresholds in pure-tone audiometry, 0.5-1% of children have difficulty understanding what they hear. An auditory processing disorder (APD) can be assumed, which should be clarified and treated. In patients with hearing loss, this must first be compensated or resolved. Only hereafter can a suspected APD be confirmed or excluded. Diagnosis of APD requires that a clear discrepancy between the child's performance in individual auditory functions and other cognitive abilities be demonstrated. Combination of therapeutical modalities is considered particularly more beneficial in APD patients than a single modality.

Treatment modalities should consider linguistic and cognitive processes (top-down), e.g., metacognitive knowledge of learning strategies or vocabulary expansion, but also address underlying auditory deficits (bottom-up). Almost 50% of children with APD also have a language development disorder requiring treatment and/or dyslexia. Therefore, each therapeutic intervention for a child with APD must be individually adapted according to the diagnosed impairments. Musical training can improve phonologic and reading abilities. Changes and adaptations in the classroom are helpful to support the weak auditory system of children with APD. Architectural planning of classrooms can be a means of ensuring that direct sound is masked by as little diffuse sound as possible. For example, acoustic ceiling tiles are suitable for reducing reverberant and diffuse sound.

Int J Pediatr Otorhinolaryngol. 2020 Mar 9;133:109984. doi: 10.1016/j.ijporl.2020.109984.

### [Hearing and speech benefits of cochlear implantation in children: A review of the literature.](#)

[Sharma SD, Cushing SL, Papsin BC, Gordon KA.](#)

**ABSTRACT:** Cochlear implantation is a safe and reliable treatment for children with severe to profound hearing loss. The primary benefit of these medical devices in children is the acquisition of hearing, which promotes development of spoken language. The present paper reviews published literature demonstrating predictive effects of a number of factors on acquisition of hearing development and speech recognition. Of the many variables that contribute to an individual child's development after implantation, age at implantation, the presence of medical comorbidities, social determinants of health, and the provision of bilateral versus unilateral hearing are those that can vary widely and have consistently shown clear impacts. Specifically, age of implantation is crucial to reduce effects of deafness on the developing auditory system and capture the remarkable plasticity of early development. Language development after cochlear implantation requires therapy emphasizing hearing and oral communication, education, and other support which can be influenced by known social determinants of health; specifically, outcomes in children decline with reductions in socioeconomic status and levels of parental education. Medical co-morbidities also slow rates of progress after cochlear implantation. On the other hand, benefits of implantation increase in children who are provided with access to hearing from both ears. In sum, cochlear implants promote development of hearing in children and the best outcomes are achieved by providing early access to sound in both ears. These benefits can be limited by known social determinants of health which restrict access to needed support and medical comorbidities which add further complexity in care and outcome.

Int J Pediatr Otorhinolaryngol. 2020 Apr;131:109881. doi: 10.1016/j.ijporl.2020.109881. Epub 2020 Jan 16.

### [Comprehensive hearing care network for early identification and intervention in children with congenital and late-onset/acquired hearing loss: 8 years' experience in Miyazaki.](#)

[Shirane M, Ganaha A, Nakashima T, Shimoara S, Yasunaga T, Ichihara S, Kageyama S, Matsuda Y, Tono T.](#)

**OBJECTIVE:** In 2010, we established the Miyazaki Comprehensive Hearing Care Network (MCHCN) for early identification and intervention in children with congenital and late-onset/acquired hearing loss with the cooperation of related administrative bodies in Miyazaki prefecture. The central roles of the MCHCN program are played by the Hearing Care Center (HCC) at the University of Miyazaki Hospital established in 2010 to facilitate audiological diagnoses, hearing aid interventions, and educational efforts, as well as linkage with the Department of Otolaryngology for surgical interventions. Herein, we aimed to present the main outcomes of the MCHCN program organized by the HCC at the University of Miyazaki Hospital.

**METHODS:** The MCHCN consists of two different networks, the Newborn Hearing Screening Network (NHSN) and the Pediatric Hearing Care Network (PHCN). All children suspected of having hearing loss by Newborn Hearing Screening (NHS) are referred to the HCC via the NHSN. In addition, children suspected of late-onset/acquired hearing loss by municipality-led health checkups, pediatricians, public health nurses, and childcare workers are referred to the HCC via the PHCN. Children who were born in Miyazaki prefecture between January 2010 and December 2017 and referred to the HCC for detailed hearing examination were included in this study.

**RESULTS:** Within the study period, 89,390 infants were born in Miyazaki prefecture, and 84,737 (94.9%) of them underwent NHS. A total of 698 infants and 182 children with suspected hearing loss were referred to the HCC via the NHSN and PHCN, respectively. Of the 880 referrals, 169 were diagnosed with hearing loss, which included 80 children with bilateral hearing loss and 89 children with unilateral hearing loss. Of the 80 children with bilateral hearing loss, 76 began wearing hearing aids and 15 had cochlear implants in the follow-up period. In children with bilateral conductive hearing loss, 4 children with bilateral middle ear anomalies underwent ossiculoplasty, following which two of these children no longer required hearing aids. Imaging assessments performed on 71 of the 89 children with unilateral hearing loss revealed that 20 of the 30 (66%) children who underwent CT exhibited ossicular anomalies and 28 out of the 48 (58%) children who underwent MRI were found to have ipsilateral cochlear nerve hypoplasia. Among the 169 children with hearing loss, no follow-up loss was observed during the period of this study.

**CONCLUSION:** The MCHCN that was organized at the initiative of the HCC at the University of Miyazaki Hospital has enabled the provision of comprehensive and continuous support, ranging from diagnosis to intervention, not

only for children with suspected hearing loss referred based on their NHS results but also for those who pass the screening. Via this system, children with late-onset/acquired hearing loss can be identified early and can receive medical interventions tailored to the cause of their hearing loss while simultaneously avoiding a loss to follow-up.

Int J Pediatr Otorhinolaryngol. 2020 Mar 6;133:109983. doi: 10.1016/j.ijporl.2020.109983.

[Endoscopic findings and long-term hearing results for pediatric unilateral conductive hearing loss.](#)  
[Silvola JT.](#)

**OBJECTIVES:** Analyze reasons for unilateral conductive hearing loss (CHL) with unknown etiology in children.

**INTRODUCTION:** Unilateral conductive hearing loss (HL) without known etiology can be undiagnosed despite of hearing screening programs. It can be difficult to find the reason for HL and to make a treatment plan. Middle ear endoscopy gives hard-evidence diagnosis and basis for an individual treatment plan.

**METHODS AND MATERIAL:** Prospective clinical follow-up study for a cohort of generally healthy elementary school age children with unilateral conductive HL with unknown etiology. The study population was 192 children, of which 46 had a HL of at least 25 dB with more than 10 dB conductive component. Mean age was 8.7 years. Preoperative tests included otomicroscopy, bone- and air-conduction audiogram, tympanometry, stapes reflex tests, Rinne and Weber test and Otoacoustic emissions. The children underwent endoscopy of the middle ear with an individual treatment plan and long-term follow-up. The aim was to explore etiology and to give a treatment plan for hearing loss. Follow-up included air- and bone conduction hearing tests annually or every other year. Mean follow-up was 5.2 years.

**RESULTS:** A clear etiological finding was found in 36 (78%) ears, stapes anomaly (23) as the most common (64%) finding. Other findings were two cholesteatomas, 2 status after trauma, 5 middle ear anomalies, 5 incus fixations and one incus erosion. Air conduction hearing improved spontaneously during follow-up in 81% (17/21, 2 dropouts) of the stapes anomaly ears (mean 11,3 dB, range 4-32 dB), and none of these ears showed hearing deterioration. In the incus fixation group, one ear showed hearing deterioration. There were no major complications for exploration, and 5 minor postoperative infections.

**CONCLUSIONS:** The most common reason for pediatric unilateral conductive hearing loss was stapes anomaly/ fixation. The HL does not deteriorate. Hearing loss in stapes anomalies shows a tendency for spontaneous recovery. Stapes surgery can be postponed or avoided.

Aust J Prim Health. 2020 Jan 20. doi: 10.1071/PY18162.

[Developmental vulnerability of Australian school-entry children with hearing loss.](#)  
[Simpson A, Šarkić B, Enticott JC, Richardson Z, Buck K.](#)

**ABSTRACT:** National data from the Australian Early Development Census (AEDC) was used to describe the sociodemographic and developmental characteristics of a cohort of Australian children entering their first year of primary school in 2012. Results, together with sociodemographic variables were reported for two groups: children with and without reported hearing loss. Data on 285232 children were analysed, with just over 1% of these children identified with hearing loss. Logistic regression analysis found that children with reported hearing loss had over double the odds than their hearing peers of being developmentally 'vulnerable' on one or more domains of the AEDC. Covariates of interest included Aboriginal and Torres Strait Islander heritage, as well as high rates of school absenteeism. Retrospective longitudinal research linking developmental outcomes with intervention efforts, such as newborn hearing screening, would be beneficial in future research.

Am J Otolaryngol. 2020 Jan 10:102398. doi: 10.1016/j.amjoto.2020.102398.

[The effect of passive smoking on the etiology of serous otitis media in children.](#)  
[Tarhun YM.](#)

**ABSTRACT:** Serous otitis media (SOM) is a disease mostly seen in the pediatric age group and characterized by serous effusion in the middle ear. The disease which is mostly silent can cause permanent hearing loss if it is not diagnosed and treated early. Passive smoking is one of the environmental factors in the etiopathology of the disease and risk factors for SOM formation in children. In our study, smoking habits of family members of 75 children with SOM and 50 healthy controls were investigated. At the end of the study, the correlation between SOM and passive smoke exposed was statistically significant in children ( $p < 0.01$ ). In this study, the effect of passive smoking, which is a preventable and controllable risk factor in the etiology of the SOM in children is emphasized.

BMC Infect Dis. 2020 Mar 17;20(1):225. doi: 10.1186/s12879-020-4950-y.

[Bacterial otitis media in sub-Saharan Africa: a systematic review and meta-analysis.](#)  
[Tesfa T, Mitiku H, Sisay M, Weldegebreal F, Ataro Z, Motbaynor B, Marami D, Teklemariam Z.](#)

**BACKGROUND:** Otitis media is inflammation of the middle ear, comprising a spectrum of diseases. It is the commonest episode of infection in children, which often occurs after an acute upper respiratory tract infection.

Otitis media is ranked as the second most important cause of hearing loss and the fifth global burden of disease with a higher incidence in developing worlds like Sub-Saharan Africa and South Asia. Therefore, this systematic review is aimed to quantitatively estimate the current status of bacterial otitis media, bacterial etiology and their susceptibility profile in sub-Saharan Africa.

**METHODS:** A literature search was conducted from major databases and indexing services including EMBASE (Ovid interface), PubMed/MEDLINE, Google Scholar, ScienceDirect, Cochrane Library, WHO African Index-Medicus and others. All studies (published and unpublished) addressing the prevalence of otitis media and clinical isolates conducted in sub-Saharan Africa were included. Format prepared in Microsoft Excel was used to extract the data and data was exported to Stata version 15 software for the analyses. Der-Simonian-Laird random-effects model at a 95% confidence level was used for pooled estimation of outcomes. The degree of heterogeneity was presented with  $I^2$  statistics. Publication bias was presented with funnel plots of standard error supplemented by Begg's and Egger's tests. The study protocol is registered on PROSPERO with reference number ID: CRD42018102485 and the published methodology is available from <http://www.crd.york.ac.uk/CRD42018102485>.

**RESULTS:** A total of 33 studies with 6034 patients were included in this study. All studies have collected ear swab/discharge samples for bacterial isolation. The pooled isolation rate of bacterial agents from the CSOM subgroup was 98%, patients with otitis media subgroup 87% and pediatric otitis media 86%. A univariate meta-regression analysis indicated the type of otitis media was a possible source of heterogeneity ( $p$ -value = 0.001). The commonest isolates were *P. aeruginosa* (23-25%), *S. aureus* (18-27%), *Proteus* species (11-19%) and *Klebsiella* species. High level of resistance was observed against Ampicillin, Amoxicillin-clavulanate, Cotrimoxazole, Amoxicillin, and Cefuroxime.

**CONCLUSION:** The analysis revealed that bacterial pathogens like *P. aeruginosa* and *S. aureus* are majorly responsible for otitis media in sub-Saharan Africa. The isolates have a high level of resistance to commonly used drugs for the management of otitis media.

Otolaryngol Head Neck Surg. 2020 Jan;162(1):114-120. doi: 10.1177/0194599819880348. Epub 2019 Oct 8.

### [Should You Follow the Better-Hearing Ear for Congenital Cytomegalovirus Infection and Isolated Sensorineural Hearing Loss?](#)

[Torrecillas V](#), [Allen CM](#), [Greene T](#), [Park A](#), [Chung W](#), [Lanzieri TM](#), [Demmler-Harrison G](#).

**OBJECTIVE:** To describe the progression of sensorineural hearing loss (SNHL) in the better- and poorer-hearing ears in children with asymptomatic congenital cytomegalovirus (CMV) infection with isolated SNHL. **STUDY**

**DESIGN:** Longitudinal prospective cohort study.

**SETTING:** Tertiary medical center.

**SUBJECTS AND METHODS:** We analyzed hearing thresholds of the better- and poorer-hearing ears of 16 CMV-infected patients with isolated congenital/early-onset or delayed-onset SNHL identified through hospital-based CMV screening of >30,000 newborns from 1982 to 1992.

**RESULTS:** By 12 months of age, 4 of 7 patients with congenital/early-onset SNHL developed worsening thresholds in the poorer-hearing ear, and 1 had an improvement in the better-hearing ear. By 18 years of age, all 7 patients had worsening thresholds in the poorer-hearing ear and 3 patients had worsening thresholds in the better-hearing ear. Hearing loss first worsened at a mean age of 2 and 6 years in the poorer- and better-hearing ears, respectively. Nine patients were diagnosed with delayed-onset SNHL (mean age of 9 years vs 12 years for the poorer- and better-hearing ears), 6 of whom had worsening thresholds in the poorer-hearing ear and 1 in both ears.

**CONCLUSION:** In most children with congenital CMV infection and isolated SNHL, the poorer-hearing ear worsened earlier and more precipitously than the better-hearing ear. This study suggests that monitoring individual hearing thresholds in both ears is important for appropriate interventions and future evaluation of efficacy of antiviral treatment.

Eur Arch Otorhinolaryngol. 2020 Mar 27. doi: 10.1007/s00405-020-05935-7.

### [Assessment of temporal processing functions in early period cochlear implantation.](#)

[Tuz D](#), [Aslan F](#), [Böke B](#), [Yücel E](#).

**PURPOSE:** The purpose of this study is to compare the temporal processing performance of children with cochlear implant (CI) according to the age of implantation and to determine their relation with auditory perception scores.

**METHODS:** In this study, 30 cochlear implant users and ten normal hearing children at 9 and 10 years were included. Children with cochlear implants are divided into two groups according to the age of implantation: group I includes participants whose implantation age is between 13 and 35 months (20 children), group II includes participants whose implantation age is between 36 and 45 months (10 children). Individuals were evaluated with random gap detection test (RGDT), duration pattern test (DPT), frequency pattern test (FPT), the Mr. Potato Head task, word recognition, and sentence recognition test.

**RESULTS:** A significant difference was found between the control and CI groups in temporal processing

performance. The temporal processing ability of CI groups was significantly worse than those of normal hearing. Although there was no significant difference among the groups with cochlear implant in terms of temporal processing performance, children who started to use CI at an earlier age showed a tendency of better performance on temporal processing tasks. There was a significant relationship between Daily Sentence Test and FPT, and the Mr. Potato Head task and FPT rev (the score calculated by accepting the reverse patterns correctly). There was a significant relationship between duration of implant use and temporal ordering performance  
**CONCLUSION:** In this study, children with CI cannot perform as well as normal-hearing peers on temporal processing tasks, even if they had started to use their CIs at an early age. It is important to evaluate temporal processing in implanted individuals and to guide auditory training considering the evaluation results.

Am J Otolaryngol. 2020 Mar - Apr;41(2):102379. doi: 10.1016/j.amjoto.2019.102379. Epub 2019 Dec 19.

### [Experience with cholesteatoma behind an intact tympanic membrane in children.](#)

[Urík M](#), [Kaliariková A](#), [Machač J](#), [Jurajda M](#).

**INTRODUCTION:** To systematically investigate all surgeries for cholesteatoma behind an intact tympanic membrane at our department. To identify predictive factors that can help the surgeon to plan surgery, surgical techniques, and follow-up treatment.

**MATERIAL AND METHODS:** This retrospective study evaluates 21 child patients, who were operated in the period 2007-2017 on for cholesteatoma behind an intact tympanic membrane.

**RESULTS:** A total of 202 primary operations were performed for cholesteatoma. In 21 cases (10,4%) there was a cholesteatoma behind an intact tympanic membrane and in 11 (5,45%) cases of it there was the congenital cholesteatoma. The most frequently affected area was the anterior-superior quadrant. The preoperative hearing loss increased significantly with disease severity (I-IV by Potsic).

**CONCLUSIONS:** The classification system according to Potsic is sufficient and fully corresponds to the surgeon's needs. It has been clearly shown that a higher CC stage is associated with worse postoperative hearing results.

Cochrane Database Syst Rev. 2020 Jan 21;1:CD010885. doi: 10.1002/14651858.CD010885.pub5.

### [Different infusion durations for preventing platinum-induced hearing loss in children with cancer.](#)

[van As JW](#), [van den Berg H](#), [van Dalen EC](#).

**BACKGROUND:** Platinum-based therapy, including cisplatin, carboplatin or oxaliplatin, or a combination of these, is used to treat a variety of paediatric malignancies. Unfortunately, one of the most important adverse effects is the occurrence of hearing loss or ototoxicity. In an effort to prevent this ototoxicity, different platinum infusion durations have been studied. This review is the third update of a previously published Cochrane Review.

**OBJECTIVES:** To assess the effects of different durations of platinum infusion to prevent hearing loss or tinnitus, or both, in children with cancer. Secondary objectives were to assess possible effects of these infusion durations on: a) anti-tumour efficacy of platinum-based therapy, b) adverse effects other than hearing loss or tinnitus, and c) quality of life.

**SEARCH METHODS:** We searched the electronic databases Cochrane Central Register of Controlled Trials (CENTRAL; the Cochrane Library 14 November 2019), MEDLINE (PubMed) (1945 to 14 November 2019) and Embase (Ovid) (1980 to 14 November 2019). In addition, we handsearched reference lists of relevant articles and we assessed the conference proceedings of the International Society for Paediatric Oncology (2009 up to and including 2019) and the American Society of Pediatric Hematology/Oncology (2014 up to and including 2019). We scanned ClinicalTrials.gov and the World Health Organization International Clinical Trials Registry Platform (WHO ICTRP; apps.who.int/trialsearch) for ongoing trials (both searched on 4 November 2019).

**SELECTION CRITERIA:** Randomised controlled trials (RCTs) or controlled clinical trials (CCTs) comparing different platinum infusion durations in children with cancer. Only the platinum infusion duration could differ between the treatment groups.

**DATA COLLECTION AND ANALYSIS:** Two review authors independently performed the study selection, 'Risk of bias' assessment and GRADE assessment of included studies, and data extraction including adverse effects. Analyses were performed according to the guidelines of the Cochrane Handbook for Systematic Reviews of Interventions.

**MAIN RESULTS:** We identified one RCT and no CCTs; in this update no additional eligible studies were identified. The RCT (total number of children = 91) evaluated the use of a continuous cisplatin infusion (N = 43) versus a one-hour bolus cisplatin infusion (N = 48) in children with neuroblastoma. For the continuous infusion, cisplatin was administered on days one to five of the cycle, but it is unclear if the infusion duration was a total of five days. Risk of bias was present. Only results from shortly after induction therapy were provided. No clear evidence of a difference in hearing loss (defined as asymptomatic and symptomatic disease combined) between the different infusion durations was identified as results were imprecise (risk ratio (RR) 1.39, 95% confidence interval (CI) 0.47 to 4.13, low-quality evidence). Although the numbers of children were not provided, it was stated that tumour response was equivalent in both treatment arms. With regard to adverse effects other than ototoxicity, we were

only able to assess toxic deaths. Again, the confidence interval of the estimated effect was too wide to exclude differences between the treatment groups (RR 1.12, 95% CI 0.07 to 17.31, low-quality evidence). No data were available for the other outcomes of interest (i.e. tinnitus, overall survival, event-free survival and quality of life) or for other (combinations of) infusion durations or other platinum analogues.

**AUTHORS' CONCLUSIONS:** Since only one eligible RCT evaluating the use of a continuous cisplatin infusion versus a one-hour bolus cisplatin infusion was found, and that had methodological limitations, no definitive conclusions can be made. It should be noted that 'no evidence of effect', as identified in this review, is not the same as 'evidence of no effect'. For other (combinations of) infusion durations and other platinum analogues no eligible studies were identified. More high-quality research is needed.

Int J Pediatr Otorhinolaryngol. 2020 Feb 4;132:109909. doi: 10.1016/j.ijporl.2020.109909.

### [Correlation of air-bone gap and size of Enlarged Vestibular Aqueduct in children.](#)

[Van Beck J](#), [Chinnadurai S](#), [Morrison AK](#), [Zuniga MG](#), [Smith B](#), [Lohse CM](#), [McCaslin D](#).

**OBJECTIVE:** Enlarged vestibular aqueduct (EVA) is an inner ear malformation that represents an important cause of pediatric hearing loss. While certain elements in the history or audiogram may suggest EVA, it is most often diagnosed using computed tomography (CT). The present investigation was conducted to determine if the size of the audiometric air-bone gap (ABG) is correlated with the size of the vestibular aqueduct in the pediatric population using three vestibular aqueduct measurements. These included the fundus, midpoint, and porous widths of the vestibular aqueduct.

**STUDY DESIGN:** This is a retrospective cohort study.

**SETTING:** This study took place at a tertiary care referral center.

**PATIENTS:** Fifty-five children (33 female; 22 male) with a confirmed diagnosis of unilateral or bilateral EVA as determined by prior imaging of the inner ear were included in the study.

**MAIN OUTCOME MEASURES:** Associations of EVA measurements with ABGs at 0.5 and 1 kHz were evaluated using Pearson correlation coefficients.

**RESULTS:** All of the correlation coefficients were positive, indicating that as EVA measurements increased so did the ABG. Only the correlation between fundus width and ABG at 1 kHz was not statistically significant.

**CONCLUSIONS:** ABGs measured during audiometric testing correlate with the size of the EVA and ABGs can be clinical predictors of the severity of the bony abnormality. These data support the third window theory of conductive hearing loss in pediatric EVA.

BMJ Case Rep. 2019 Dec 1;12(11). pii: e231978. doi: 10.1136/bcr-2019-231978.

### [Neonatal cholestasis, hyperferritinemia, hypoglycemia and deafness: a diagnostic challenge.](#)

[van Westering-Kroon E](#), [Heijligers M](#), [Hütten MC](#).

**ABSTRACT:** Neonatal conjugated hyperbilirubinemia is a diagnostic challenge. A full term, small for gestational age boy presented with cholestasis, hypoglycemia, hyperferritinemia and severe bilateral deafness. Diagnostic work-up revealed two hereditary diseases: alpha-1-antitrypsin deficiency (PI\*ZZ genotype) and autosomal recessive deafness type 3 (compound heterozygous MYO15A gene mutation). In addition, we found late hypoglycemia on full enteral feeding which complicated this case. Hyperferritinemia is an uncommon finding in newborn cholestasis without liver failure.

Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2020 Feb;34(2):113-118. doi: 10.13201/j.issn.1001-1781.2020.02.004. [Article in Chinese]

### [Analysis of genotypes and hearing phenotypes of mutation infants with deafness.](#)

[Wang X](#), [Zhao X](#), [Huang L](#), [Wen C](#), [Wang X](#), [Cheng X](#).

**OBJECTIVE:** The aim of this study is to explore the genotype and hearing phenotype of deaf infants with mutation of *GJB2* gene.

**METHOD:** Subjects were 121 infants with *GJB2* gene mutations who were treated in the Children's Hearing Diagnosis Center of Beijing Tongren hospital. All subjects were accepted to undertake the universal newborns hearing screening (UNHS) and series of objective audiometry, including auditory brainstem response, distortion product otoacoustic emission, auditory steady-state response and other audiological tests. All subjects were screened for nine pathogenic variants in four genes or all exons of the *GJB2* gene, and then were diagnosed as infants with *GJB2* gene mutations. Initially, analyzing their genotypes and hearing phenotypes generally. Then, the subjects were divided into two groups according to the genotypes: T/T group (truncated/truncated mutations, 89 cases) and T/NT group (truncated/non-truncated mutations, 32 cases). Chi-square test was used to analyze the results of UNHS, hearing degree, audiogram patterns and symmetry/asymmetry of binaural hearing phenotype. Eventually, analyzing the results of UNHS.

**RESULT:** The most common truncated mutation was c.235delC (64.88%, 157/242 and the most common non-

truncated mutation was c.109G>A (11.16%, 27/242). The homozygous mutation of c.235delC/c.235delC was the dominant in T/T group (38.84%, 47/121), and the compound heterozygous mutation of c.235delC/c.109G>A was the dominant in T/NT group (18.18%, 22/121). 81.82% (99/121) of subjects failed in UNHS, including 74.38% (90/121) with bilateral reference, 7.44% (9/121) with a single pass. The refer rate of UNHS of group T/T and T/NT were 86.52% (77/89) and 68.75%, respectively. There was a statistically significant difference between the two groups ( $P<0.05$ ). 85.95% (104/121) of subjects were diagnosed as hearing loss and 14.05% (17/121) of subjects were diagnosed as normal hearing. The degree of hearing loss: profound, severe, moderate and mild were 31.40% (38/121), 19.01% (23/121), 24.79% (30/121) and 10.74% (13/121), respectively. There were no subjects with normal hearing in T/T group and individuals with severe and profound hearing loss accounted for the highest proportion (65.17%, 58/89), while in T/NT group, normal hearing accounted for 53.13% (17/32) and mild and moderate hearing loss accounted for the highest proportion (37.5%, 12/32). There was a statistically significant difference between the two groups ( $P<0.05$ ). Of 104 patients (208 ears with hearing loss), the audiogram patterns: flat, descending, ascending, residual, Valley and other types were 49.03% (102/208), 12.02% (25/208), 8.65% (18/208), 7.69% (16/204), 3.36% (7/204) and 19.23% (40/204), respectively. The two most common types in T/T group were flat (47.19%, 84/178) and other types (20.22%, 36/178), while in T/NT group were flat (60.00%, 18/30) and ascending (20.00%, 6/30). There was a statistically significant difference between the two groups ( $P<0.05$ ). There were 50 cases (48.07% with symmetrical hearing phenotype) and 54 cases (51.93%) with asymmetrical hearing phenotype. Asymmetry was predominant in T/T group (53.93%, 48/89), and symmetry was predominant in T/NT group (60.00%, 9/15). There was no statistically significant difference between the two groups ( $P>0.05$ ).

**CONCLUSION:** In this study, c.235delC/c.235delC homozygous mutation was dominant in T/T group and c.235delC/c.109G>A heterozygous mutation was dominant in T/NT Group. The hearing phenotypes in T/T group were mostly bilateral asymmetric severe hearing loss, and those in T/NT Group were bilateral symmetric mild to moderate hearing loss, special attention should be paid to the audiological characteristics of different genotypes.

Otolaryngol Head Neck Surg. 2020 Apr 14;194599820915741. doi: 10.1177/0194599820915741.

### [Cognitive and Behavioral Functioning in Hearing-Impaired Children with and without Language Delay.](#)

[Williams A](#), [Pulsifer M](#), [Tissera K](#), [Mankarious LA](#).

**ABSTRACT:** Poor language development in patients with sensorineural hearing loss (SNHL) may be related to an auditory deficit and/or other neurologic condition that influences the ability to communicate. A retrospective chart review of children (mean age = 4.0 years) with congenital, bilateral SNHL was performed to assess for linguistic and nonlinguistic neurodevelopmental differences between those who were language-impaired (LI) versus non-language-impaired (NLI). Language, neurodevelopmental functioning, and behavior were assessed. Twenty-two patients were identified: 12 were LI and 10 were NLI. Average pure-tone thresholds and nonverbal intelligence were not different between the language groups, but the LI group demonstrated significantly lower median overall adaptive skills, personal living skills, and motor skills. Behavioral dysregulation was significantly higher in the LI versus NLI group (58% vs 10%;  $P = .031$ ), although the median neurodevelopmental scores did not differ significantly. These findings introduce the possibility that nonlinguistic processing deficit(s) may be confounding the ability to develop language.

Zhonghua Yi Xue Yi Chuan Xue Za Zhi. 2020 Mar 10;37(3):269-276. doi: 10.3760/cma.j.issn.1003-9406.2020.03.008. [Article in Chinese]

### [Clinical practice guidelines for hereditary non-syndromic deafness.](#)

[Writing Group For Practice Guidelines For Diagnosis And Treatment Of Genetic Diseases Medical Genetics Branch Of Chinese Medical Association](#), [Yuan H](#), [Dai P](#), [Liu Y](#), [Yang T](#).

**ABSTRACT:** Genetic factors are a common cause for non-syndromic hearing loss (NSHL). Along with the development and maturity of molecular techniques, genetic diagnosis and counseling is increasingly affecting the clinical practice of NSHL. Newborn hearing screening has facilitated early detection of affected children, whilst genetic screening has enabled identification of the cause of NSHL, and genetic diagnosis and consultation can promote early intervention of deafness. So far 110 pathogenic genes of NSHL have been discovered, though there are still many challenges lying in its clinical identification. The development of genetic counseling and prenatal diagnosis has put forward greater requirements for genetic testing and data interpretation. This guideline has summarized the incidence, mutational spectrum, inheritance mode, pathogenesis, clinical manifestation, genotype - phenotype correlation, genetic testing, treatment and intervention, as well as risk assessment for NSHL, with an aim to provide a reference for genetic consultants, clinical otologists and professionals engaged in genetic testing.

## [Contribution of Congenital Cytomegalovirus Infection to Permanent Hearing Loss in a Highly Seropositive Population: The Brazilian Cytomegalovirus Hearing and Maternal Secondary Infection Study.](#)

[Yamamoto AY](#), [Anastasio ART](#), [Massuda ET](#), [Isaac ML](#), [Manfredi AKS](#), [Cavalcante JMS](#), [Carnevale-Silva A](#), [Fowler KB](#), [Boppana SB](#), [Britt WJ](#), [Mussi-Pinhata MM](#).

**BACKGROUND:** The exact contribution of congenital cytomegalovirus infection (cCMVI) to permanent hearing loss (HL) in highly seropositive populations is unknown. We determined the contribution of cCMVI to HL and estimated the effectiveness of newborn hearing screening (HS) in identifying neonates with CMV-related HL.

**METHODS:** A total of 11 900 neonates born from a population with  $\geq 97\%$  maternal seroprevalence were screened for cCMVI and HL. cCMVI was confirmed by detection of CMV-DNA in saliva and urine at age  $< 3$  weeks.

**RESULTS:** Overall, 68 (0.6%; 95% confidence interval [CI], 0.4-0.7) neonates were identified with cCMVI. Of the 91 (0.8%) newborns who failed the HS, 24 (26.4%) were confirmed with HL, including 7 (29.2%; 95% CI, 17.2-59.3) with cCMVI. Another newborn with cCMVI passed the HS but was confirmed with HL at age 21 days. Of the 62 neonates with cCMVI who underwent a complete hearing evaluation, 8 (12.9%; 95% CI, 6.7-23.4) had HL and most (7/8; 87.5%; 95% CI, 46.6-99.7) were identified by HS. The rate of CMV-related HL was 8 per 11 887 neonates (0.7 per 1000 live births). The prevalence ratio of HL among neonates with cCMVI compared to CMV-uninfected neonates was 89.5 (95% CI, 39.7-202.0). No late-onset cCMVI-related HL was detected during a median follow-up of 36 months.

**CONCLUSIONS:** cCMVI is an important cause of HL in childhood in all settings. Integrating targeted cCMVI screening among neonates who fail a HS could be a reasonable, cost-effective strategy to identify newborns with early-onset cCMVI-related HL.

## [Cost-effectiveness of School Hearing Screening Programs: A Scoping Review.](#)

[Yong M](#), [Liang J](#), [Ballreich J](#), [Lea J](#), [Westerberg BD](#), [Emmett SD](#).

**OBJECTIVE:** School hearing screening is a public health intervention that can improve care for children who experience hearing loss that is not detected on or develops after newborn screening. However, implementation of school hearing screening is sporadic and supported by mixed evidence to its economic benefit. This scoping review provides a summary of all published cost-effectiveness studies regarding school hearing screening programs globally. At the time of this review, there were no previously published reviews of a similar nature.

**DATA SOURCES:** A structured search was applied to 4 databases: PubMed (Medline), Embase, CINAHL, and Cochrane Library.

**REVIEW METHODS:** The database search was carried out by 2 independent researchers, and results were reported in accordance with the PRISMA-ScR checklist and the JBI methodology for scoping reviews. Studies that included a cost analysis of screening programs for school-aged children in the school environment were eligible for inclusion. Studies that involved evaluations of only neonatal or preschool programs were excluded.

**RESULTS:** Four of the 5 studies that conducted a cost-effectiveness analysis reported that school hearing screening was cost-effective through the calculation of incremental cost-effectiveness ratios (ICERs) via either quality- or disability-adjusted life years. One study reported that a new school hearing screening program dominated the existing program; 2 studies reported ICERs ranging from 1079 to 4304 international dollars; and 1 study reported an ICER of £2445. One study reported that school-entry hearing screening was not cost-effective versus no screening.

**CONCLUSION:** The majority of studies concluded that school hearing screening was cost-effective. However, significant differences in methodology and region-specific estimates of model inputs limit the generalizability of these findings.

## [Parents' satisfaction with a trial of a newborn hearing screening programme in Jordan.](#)

[Zaitoun M](#), [Nuseir A](#).

**OBJECTIVE:** This study examines parents' satisfaction level toward a trial of a newborn hearing screening programme (NHSP) that was applied in King Abdullah II University Hospital (KAUH) in Jordan over one year. This is the first study that investigated parents' satisfaction toward a hearing screening programme in the Arab countries, and the results will improve any future screening programmes in the Arabian region.

**METHOD:** The main tool for this study was a questionnaire that was translated and modified from the original version of the Parental Satisfaction with the Newborn Hearing Screening Programme (PSQ-NHSPs1). The questionnaire consisted of 19 items covering five main aspects of the NHSP. The parents' responses were not anonymously given where the parents whose children had undergone the hearing screening were contacted by



phone using the data record of the hospital.

**RESULTS:** The majority of the parents were very satisfied with the programme overall and showed great support and appreciation for the effort in testing their babies and increasing their awareness. The satisfaction levels varied among the specific aspects of the programme. Good portion of the parents did not receive the brochure containing information about the screening, and almost half of them did not know the results of the hearing screening.

**CONCLUSION:** Parents were overall satisfied with neonatal hearing screening programme that was conducted at KAUH. However, parents were less satisfied with information related to the test procedure and results. Parents' responses in this study could be used to improve any future hearing screening program in Jordan or in the Arab countries.