

Others' Publications about EHDl: May 2019 through November 2019

The *Journal of Early Hearing Detection and Intervention (JEHDI)* publishes peer-reviewed articles that describe 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. The aim of the journal is to improve Early Hearing Detection and Intervention (EHDl) systems.

Although *JEHDI* is the only journal that focuses exclusively on improving EHDl systems, many other journals include articles relevant to *JEHDI*'s aim as a part their journal's broader focus. To help *JEHDI* readers stay up-to-date about current research and practices related to improving EHDl programs, we provide titles and abstracts of recent publications that are relevant to improving EHDl programs. Titles of all articles are hyperlinked to the source.

The EHDl-relevant articles referred to in the following abstracts are from all over the world – demonstrating the breadth and depth of EHDl programs. Interestingly, authors of these articles are based in 37 different countries and almost 50% are from low and middle income countries. Many of the abstracts listed below focus on the basic components of EHDl systems (e.g., screening, diagnosis, early intervention). This suggests that there are still areas in the basic EHDl system that need to be improved, but many other articles are breaking new ground and suggesting creative innovations. For example:

- Dai et al. reported the results of a study done with 180,469 infants born in Beijing, who were screened for hearing loss using transient evoked otoacoustic emission and automated auditory brainstem response. Dried blood spots for each infant were also collected and analyzed to screen for mutations related to hearing loss in nine variants of four genes (GJB2, SLC26A4, mtDNA 12S rRNA, and GJB3) using a DNA microarray platform. The authors concluded that concurrent screening for hearing loss and genetic mutations provides a more comprehensive approach for management of congenital deafness.
- Davidson et al., evaluated the optimal level and duration of acoustic experience for facilitating language development of pediatric cochlear implant (CI) recipients to determine if there is an optimal duration of hearing aid use and unaided threshold levels that should be considered before proceeding to bilateral CIs. Based on analyses of 117 pediatric CI recipients (ages 5–9 years), they concluded that for children with the most profound losses, early bilateral CIs provide the greatest opportunity for developing good spoken language skills. For children with moderate-to-severe losses, however, a prescribed period of bimodal use may be more advantageous for developing good spoken language skills.
- Gruber et al. conducted ophthalmologic examinations of 91 young children diagnosed with congenital sensorineural hearing loss in New Zealand and found that 21% had ophthalmic conditions that needed treatment. They concluded that ophthalmic assessment of babies and children with congenital sensorineural hearing loss allows early diagnosis and treatment of coexisting ophthalmic and hearing conditions.
- Huber and Havas from Austria did a systematic review of 30 studies that addressed the quality of life of children and adolescents with cochlear implants and concluded that even though many of the studies had methodological weaknesses, difficulties with speech recognition in noise may be a burden for the quality of life for these children and adolescents. The authors concluded that further studies addressing this topic are urgently needed. Interestingly, a number of other studies published during this time period also addressed Quality of Life issues (see abstracts below for Osborne et al.; Ronner et al.; Suneel et al.; Urik et al.; van der Straaten et al.; and Wang et al.)
- Krishnan et al. examined whether providing expectant mothers in the United States with information related to the newborn hearing screening process would increase their satisfaction with the screening program. Their results suggested that educating obstetricians and nurses about newborn hearing screening and having them provide information to expectant mothers, as well as having mothers present for the screening increases parental satisfaction and may therefore benefit early hearing detection and intervention programs.

- Skarżyński et al. conducted a population-based epidemiological study to investigate the prevalence of hearing loss among Polish school-aged children from rural areas. Based on pure-tone assessments of 67,416 children with a mean age of 8.65 years, they concluded that 16.4% of the sample had pure-tone average hearing losses higher than 20 dB in one or both ears. According to the authors, the study demonstrated the strong need for systematic monitoring of hearing status among children and increasing awareness of parents and educators of the significance of hearing loss, including unilateral and mild hearing loss.
- Xiao et al. assessed the efficacy of a tablet-based hearing game as a screening instrument for pediatric hearing loss. Five hundred sixteen children (ages 3–13) completed the tablet game and were also assessed with standard audiometry to assess hearing status. The overall rate of sensorineural hearing loss in the group was 4.8% and the sensitivity and specificity of the tablet game compared to the audiometric assessment was 91% and 73.5% respectively for ages 4 and older. The authors concluded that the tablet game audiometry was a reliable screening method for school-age children.

Abstracts for all 148 articles are listed below.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109659. doi: 10.1016/j.ijporl.2019.109659. Epub 2019 Aug 26.

High-frequency otoacoustic emissions in universal newborn hearing screening.

Akinpelu OV, Funnell WRJ, Daniel SJ.

OBJECTIVE: Distortion-product otoacoustic emissions (DPOAEs) are currently used in many newborn hearing screening programs as the initial hearing test, typically testing frequencies between 1 and 4 or 6 kHz, but they have been associated with high false-positive rates. The objective was to investigate the possible benefit of high-frequency DPOAEs for reducing false-positive rates.

METHODS: 255 healthy newborns (138 males and 117 females) undergoing conventional hearing screening based on DPOAE and automated auditory brainstem response (AABR) testing were recruited. High-frequency DPOAE amplitudes, noise floors and signal-to-noise ratios (SNRs) were measured for f2 frequencies up to 12 kHz.

RESULTS: Of the 255 newborns who participated in this study, 23 (9%) failed the conventional DPOAE test but passed the AABR test, and 8 (3%) failed both tests. For an SNR threshold of 6 dB, high-frequency DPOAE tests at f2=4, 6, 8 and 10 kHz resulted in a reduction in the false-positive rate from 9% to 0.4%, or to zero if only three of the four frequencies were required to exceed the threshold. SNRs were lower in newborns with birth weights greater than 4000 g; lower at 2 kHz in newborns with a gestational age of 41 weeks; slightly higher in vaginally-delivered newborns; and higher at 2 kHz with increasing age in the group that failed the conventional DPOAE test but passed AABR.

CONCLUSION: High-frequency DPOAEs resulted in a reduction in the DPOAE failure rate and the false-positive rate. These findings may be helpful in universal newborn hearing screening programs.

Int J Pediatr Otorhinolaryngol. 2019 Nov 18;129:109784. doi: 10.1016/j.ijporl.2019.109784. [Epub ahead of print]

Relation of public health staffing to follow-up after newborn hearing screening in three health districts in Georgia, 2009-2015.

Al-Mulki K, Todd NW.

OBJECTIVE: To describe the association, or non-association, of public health district staffing (specifically, Early Hearing Detection and Intervention [EHDI] coordinator/navigator) and loss to follow-up in newborns who did not pass hearing screening in selected public health districts in Georgia, USA.

METHODS: By Freedom of Information request, data regarding newborn hearing screening and loss to follow-up for diagnostic testing and staffing were acquired for three districts in Georgia for six years. The districts were chosen because their coordinator/navigator positions were unfilled at times.

RESULTS: Lapses in staffing of the district EHDI coordinator/navigator position aligned temporally with decreased follow-up. Aggregate three district data showed that follow-up rates in quarter-years with a fulltime navigator were higher than quarter-years without a full-time navigator ($p < .001$).

CONCLUSION: Lapses in staffing dedicated to EHDI navigation-coordination correlated with poorer follow-up after not passing newborn hearing screening.

Audiology and Speech-Language Pathology Simulation Training on the 1-3-6 Early Hearing Detection and Intervention Timeline.

Alanazi AA, Nicholson N.

PURPOSE: This study aimed to find out more about the effect of the use of the 1-3-6 early hearing detection and intervention (EHDI) timeline and interprofessional simulation training on audiology and speech-language pathology graduate students' confidence level of knowledge and skills.

METHOD: A 1-group pretest-posttest quasi-experimental study design was used to assess self-ratings of confidence in knowledge and skills. The mean age for 50 graduate students participating in this study was 25.32 years (SD = ± 3.77 , range: 22-37 years). Participants completed a pre- and postevent questionnaire in which they rated their level of confidence for specific knowledge and skills. Three case scenarios represented by standardized parents were developed, and 8 students volunteered to participate in implementing the 1-3-6 EHDI timeline, whereas others participated as observers. All participants participated in the briefing and debriefing sessions immediately before and after each scenario. Participants were asked to rate their readiness for interprofessional education/practice and their satisfaction of the educational experience after the last case scenario.

RESULTS: Overall, the pre- and postsimulation event questionnaire revealed a significant improvement in the participants' self-rated confidence levels in knowledge and skills. The mean difference between pre- and postevent scores was 0.78 ($p < .01$). The mean interprofessional learning was 2.13 (range: 1.16-2.57, SD = ± 0.24) based on a Likert scale, where 1 = strongly agree and 5 = strongly disagree. The mean satisfaction level was 4.37 (range: 3.94-4.72, SD = ± 0.24) based on a Likert scale, where 1 = not satisfied and 5 = very satisfied.

CONCLUSIONS: The results demonstrated the value of using interprofessional simulation training among audiology and speech-language pathology students to improve their confidence in knowledge and skills. The curriculum developed in this study for the 1-3-6 EHDI timeline provides resources for educators in both professions and other related professions.

Int J Pediatr Otorhinolaryngol. 2019 Oct 11;128:109717. doi: 10.1016/j.ijporl.2019.109717. [Epub ahead of print]

Parents' views about factors facilitating their involvement in the oral early intervention services provided for their children with hearing loss in Kuwait.

Alduhaim A, Purcell A, Cumming S, Doble M.

OBJECTIVES: Developed countries (such as the United States of America, the United Kingdom, and Australia) provide high-quality services to children with hearing loss and their parents, including universal newborn hearing screening, early fitting of hearing devices and access to high quality early, aural intervention. In contrast, many developing countries lack the resources to provide these services. Research evidence suggests that one way to compensate for delays in identification and rehabilitation for children with hearing loss is to involve the family in the early intervention process. However, evidence is deficient around the facilitation of parental involvement in early intervention for families from developing countries. The aim of this study was to investigate the perception of parents from a developing country who have a child with hearing loss to discover the factors that may influence their involvement in early intervention, identify facilitators/barriers for their involvement, and identify the specific needs of involved parents.

METHOD: A qualitative collective case study methodology was used in the form of semi-structured interviews with twenty-one participants.

RESULTS: Two global themes emerged. Firstly, factors influencing parental involvement. This included five subthemes: better communication as the key to building rapport and providing support; disappointment with the service; physical and human resources make a difference; consideration of life beyond the habilitation sessions; and my child matters the most. The second global theme-parental needs-included four subthemes: building trust; ensuring generalization; improving child outcomes; and managing needs of parents versus services. All the discussed themes are supported by quotations from the interviews.

CONCLUSION: The findings suggest that parents of children with hearing loss in developing countries perceive the services provided for their children to be disjointed and not well coordinated. Based on the parent perceptions a set of guidelines that provide a scaffold for service providers working with children with hearing loss in developing countries is outlined. The guidelines are designed to assist services for children with hearing loss in developing countries improve the delivery of their services and increase family involvement, which in turn; will assist in improving their child and family related outcomes.

Age of identification of sensorineural hearing loss and Characteristics of affected children: Findings from two cross-sectional studies in Saudi Arabia.

[Alkahtani R](#), [Rowan D](#), [Kattan N](#), [Alwan NA](#).

OBJECTIVES: To identify the average age of identification (AOI) and characteristics of Saudi children with sensorineural hearing loss (SNHL).

METHODS: Two cross-sectional studies were undertaken. Study A: the medical records of 1166 children aged 0-10 years old who visited the audiology clinics in four hospitals in Riyadh and Dammam during 2015 were reviewed. Study B: 174 carers of children aged 0-12 years who visited the audiology clinics in four hospitals in Riyadh during a three-month period were surveyed.

RESULTS: The mean AOI with SNHL in children was 3.2 years (SD=2.5 years) and 3.1 years (SD=2.6 years) with 14% and 16% not identified until after primary school age for Studies A and B, respectively. The presence of SNHL was positively associated with parental consanguinity, positive family history of SNHL, history of chemotherapy treatment, brain pathology and prior parental concern regarding their child's hearing.

CONCLUSION: AOI of SNHL among Saudi children is deemed high in relation to the likely age of onset, with about 15 in 100 children identified after school age. Childhood hearing screening programmes (at birth and at school entry) should be considered in order to intervene earlier.

Pediatr Infect Dis J. 2019 Nov 4. doi: 10.1097/INF.0000000000002510. [Epub ahead of print]

Cytomegalovirus Seroprevalence and Birth Prevalence of Congenital CMV Infection in Bosnia and Herzegovina: A Single-Center Experience.

[Arapović J](#), [Rajić B](#), [Pati S](#), [Brizić I](#), [Azinović I](#), [Šušak B](#), [Ostojić M](#), [Tutiš B](#), [Raguž AB](#), [Tomić V](#), [Jonjić S](#), [Boppana S](#).

BACKGROUND: Congenital cytomegalovirus infection (cCMV) is a leading cause of sensorineural hearing loss (SNHL) and neurodevelopmental disabilities in developed countries. Although high cCMV rates have been reported in populations with high seroprevalence, the cCMV prevalence in low/middle-income countries in Europe has not been defined.

OBJECTIVE: To determine cytomegalovirus (CMV) seroprevalence and the cCMV prevalence in Bosnia and Herzegovina.

METHODS: Between March 2010 and February 2019, 5222 sera samples from patients seen at the University Clinical Hospital Mostar were tested for CMV IgG. The cord blood samples collected from 2091 infants between July 2011 and January 2013 were analyzed for CMV IgG and CMV DNA. The cCMV prevalence was determined by testing saliva swabs from 1293 infants between November 2015 and October 2016.

RESULTS: The overall CMV IgG prevalence was 81.4% (95% confidence interval: 0.8-0.82). Significantly higher prevalence was observed among females (84.9%) than in males (77.0%), and the rate increased from 50.8% in the 1 to 5 years group to 97.7% in the group > 65 years old. Most cord blood samples (2091/1925, 92.1%) were CMV IgG positive, and 2 (0.1%) were CMV DNA positive. Of the 1293 saliva swabs, 8 (0.62%; 95% confidence interval: 0.3-1.2) were CMV positive. All 8 infected infants had asymptomatic cCMV, and none had SNHL at 18 months of age.

CONCLUSIONS: In a highly CMV seropositive population, the prevalence of cCMV was lower compared to that reported from other low/middle-income countries populations. None of the infected infants had symptomatic infection or SNHL at 18 months.

HNO. 2019 Nov;67(11):843-854. doi: 10.1007/s00106-019-0676-9.

[Objective frequency-specific measurement of hearing threshold using narrow-band chirp stimuli with level-adaptive simultaneous masking].

[Baljić I](#), [Walger M](#).

BACKGROUND: In the past, various simulation and measurement paradigms have been introduced and evaluated in order to improve frequency-specific measurement of the hearing threshold using early auditory evoked potentials (EAEP). A promising approach for improvement of detection of stimulus response is the usage of frequency-modulated chirp signals, which optimize the temporal synchrony of neuronal responses along a region of the basilar membrane.

AIM OF THE STUDY: This study validated the performance of three generated narrow-band chirp stimuli in combination with a level-adaptive simultaneous masker on a collective of normally hearing subjects.

MATERIAL AND METHODS: In this study 25 normal hearing subjects took part after undergoing pure tone audiometry as well as an objective estimation of the auditory threshold using low, middle and high chirp stimuli.

The characteristic EAEP parameters were visually identified before statistical analysis. The characteristic latency level function was conducted using measurements within a stimulus level range from 80 to 0 dB HL. Afterwards a comparison of objectively verified auditory threshold and subjective auditory threshold was conducted.

RESULTS: All objectively determined thresholds of the frequency-specific evoked EAEP were on average below 10 dB HL: low chirp at 8.2 dB HL, middle chirp at 5.8 dB HL and high chirp at 5.4 dB HL. The mean difference compared to subjectively determined auditory thresholds at all frequencies was below 3 dB and was not significant.

CONCLUSION: Brainstem evoked response audiometry (BERA) using a band-limited and level-specific masked chirp stimulus is an efficient method for the determination of frequency-specific excitation thresholds in the clinical routine. The small, insignificant difference compared to the subjectively determined auditory thresholds makes usage of correction factors mostly redundant. Confirming the study results concerning low chirp stimuli so far, the low chirp BERA currently seems to be the method of choice for estimation of auditory threshold at low frequency ranges around 500 Hz.

J Binocul Vis Ocul Motil. 2019 Jul-Sep;69(3):126-130. doi: 10.1080/2576117X.2019.1625629. Epub 2019 Jun 17.

[Ophthalmologic Abnormalities in Children with Congenital Sensorineural Hearing Loss.](#)

Batson S, Kelly K, Morrison D, Virgin F.

OBJECTIVE: To review a single center experience with the diagnosis of visual impairment in patients with sensorineural hearing loss (SNHL) and propose a diagnostic algorithm.

STUDY DESIGN: Retrospective study of patients with SNHL who were diagnosed with ophthalmologic abnormalities in the course of evaluation.

SETTING: University children's hospital and university-associated eye institute.

SUBJECTS AND METHODS: Children with the diagnosis of sensorineural hearing loss aged 0-18 who received a formal ophthalmology examination between the dates of December 2000-December 2016 were included for analysis. Children were identified using ICD-9 and ICD-10 billing codes. Primary measures included diagnosis of SNHL, ophthalmologic diagnoses, and referral source.

RESULTS: Two hundred and sixty-nine patients with SNHL met inclusion criteria. One hundred and thirty-one (48.5%) of these patients had an ophthalmic abnormality. When evaluating referral source, patients referred by a pediatrician following failed vision screen or visual complaint were more likely to have an ophthalmologic finding (61%, n = 147) when compared to referral by an otolaryngologist following diagnosis of SNHL (9.6%, n = 73). Seventeen of the 131 (13%) patients with at least one ophthalmic abnormality had an abnormality that was deemed unlikely to be detected by routine screening.

CONCLUSION: Our study agreed with previously published works that there is a high rate of ophthalmic abnormalities in patients with SNHL. Evaluation of referral source for ophthalmology evaluation suggests that routine referral by otolaryngologists in patients with SNHL may not be an efficient means of identifying patients with treatable ophthalmic disease. Reliance on school and office screenings to detect ophthalmic abnormalities, prior to referral, is likely a more efficient model, even among patients with SNHL.

Int J Pediatr Otorhinolaryngol. 2019 Nov;126:109635. doi: 10.1016/j.ijporl.2019.109635. Epub 2019 Aug 10.

[Parent-perceived challenges related to the pediatric cochlear implantation process and support services received in South Africa.](#)

Bhamjee A, Roux TL, Schlemmer K, Perold J, Cass N, Schroeder K, Schlesinger D, Ceronio D, Vinck B.

OBJECTIVE: To determine and describe parent-perceived challenges related to the pediatric cochlear implantation process and support services received.

METHOD: A multicenter survey study across six cochlear implant (CI) programs in South Africa (SA) was conducted. The study sample included 82 parents of pediatric (≤ 18 years) CI recipients with at least 12 months CI experience. A self-administered questionnaire was developed for the purpose of this study, exploring parental challenges regarding the CI process, education of their implanted children and the support services received.

RESULTS: The financial implications of cochlear implantation, including CI device maintenance, were identified by parents as the most prominent challenge. Financing issues were the highest scoring reason that attributed to the delay between diagnosis of hearing loss and cochlear implantation, as well as the greatest barrier to bilateral implantation. Parent-perceived educational challenges included finding adequate educational settings specific to the individual needs of their child and a shortage of trained teachers equipped to support children with CIs. The presence of one/more additional developmental conditions and grade repetition were associated with more pronounced parent-perceived educational challenges. Parents considered speech-language therapy as the most critical support service for their implanted children to achieve optimal outcomes, while parent guidance was indicated to be the most critical support service required for parents of pediatric CI recipients.

CONCLUSION: A greater understanding of parent-perceived challenges will guide CI professionals to promote optimal outcomes, evidence-based service delivery and on-going support to pediatric CI recipients and their families. Study results imply a call for action regarding financial and educational support for pediatric CI recipients in SA.

Indian J Otolaryngol Head Neck Surg. 2019 Nov;71(Suppl 2):1296-1299. doi: 10.1007/s12070-018-1352-1. Epub 2018 Apr 17.

Newborn Hearing Screening: Time to Act!

Bishnoi R, Baghel S, Agarwal S, Sharma S.

The present study was carried out with the aim of assessing the outcomes of otoacoustic hearing screening in newborns coupled with the three stage protocol. It was a hospital based observational study which was conducted over a time period of twelve months at a tertiary care institute to screen 2000 live neonates for congenital hearing impairment using OAE, followed up by tympanometry and BERA, if required. 2000 neonates were screened for hearing impairment. 406 were in high risk group and the rest in non-high risk group. Seven neonates had absent V wave on BERA. Five of them were high risk babies and the rest two were non-high risk ones. In order to ensure that early detection and effective intervention are possible for all neonates with hearing impairment, UNHS should be performed. Three stage UNHS protocol using OAE and BERA showed that the implementation of UNHS for congenital childhood hearing loss for all neonates would be beneficial.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109660. doi: 10.1016/j.ijporl.2019.109660. Epub 2019 Aug 25.

Effects of type 1 diabetes mellitus on efferent auditory system in children and adolescents.

Braite N, da Cruz Fernandes L, Rissatto Lago MR, de Aragão Dantas Alves C.

AIM: To investigate whether type 1 diabetes mellitus (T1DM) could affect the efferent auditory system by analyzing the relationship between the activation of the medial olivocochlear reflex with disease duration, metabolic control and age at time of diagnosis.

METHOD: A total of 101 children and adolescents were evaluated. They were divided into two groups: 50 with T1DM and 51 without the disease. The participants answered a structured questionnaire containing questions about auditory complaints and were evaluated for tonal audiometry, tympanometry, acoustic reflex, otoacoustic emission by distortion product to evaluate the inhibitory effect of medial olivocochlear reflex (MOC).

RESULTS: The participants with T1DM presented changed AR (increased or absent) at all the frequencies in both ears ($p < 0.05$) when compared with the group without the disease. No differences were found between the DPOAE amplitudes of the individuals with and without T1DM, in both ears at all the frequencies. There were significant differences in the activation of the MOC reflex between the groups with and without T1DM, the participants with T1DM presented the absence of the inhibitory effect of the DPOAE in the right and left ears, in the frequencies of 4000 Hz ($p = 0.035/0.002$ respectively) and 6000 Hz ($p = 0.033/0.031$ respectively) and 8000 Hz ($p = 0.007/0.001$ respectively) when compared to the healthy group. Significant differences were also observed between the groups with and without T1DM ($p < 0.05$) for self-reported complaints of tinnitus, difficulties with the perception of speech when there was noise and distraction with noise. No association was found between the hearing complaints and the audiological measurements obtained and disease time, metabolic control and age at the time of diagnosis.

CONCLUSION: The findings suggest the presence of early auditory dysfunction of the efferent pathway in patients with T1DM.

PLoS One. 2019 Jul 11;14(7):e0219600. doi: 10.1371/journal.pone.0219600. eCollection 2019.

Prevalence of permanent childhood hearing loss detected at the universal newborn hearing screen: Systematic review and meta-analysis.

Butcher E, Dezateux C, Cortina-Borja M, Knowles RL.

CONTEXT: Permanent childhood hearing loss (PCHL) can affect speech, language, and wider outcomes. Adverse effects are mitigated through universal newborn hearing screening (UNHS) and early intervention.

OBJECTIVE: We undertook a systematic review and meta-analysis to estimate prevalence of UNHS-detected PCHL (bilateral loss ≥ 26 dB HL) and its variation by admission to neonatal intensive care unit (NICU). A secondary objective was to report UNHS programme performance (PROSPERO: CRD42016051267).

DATA SOURCES: Multiple electronic databases were interrogated in January 2017, with further reports identified from article citations and unpublished literature (November 2017).

STUDY SELECTION: UNHS reports from very highly-developed (VHD) countries with relevant prevalence and performance data; no language or date restrictions.

DATA EXTRACTION: Three reviewers independently extracted data and assessed quality.

RESULTS: We identified 41 eligible reports from 32 study populations (1799863 screened infants) in 6195 non-duplicate references. Pooled UNHS-detected PCHL prevalence was 1.1 per 1000 screened children (95% confidence interval [CI]: 0.9, 1.3; $I^2 = 89.2\%$). This was 6.9 times (95% CI: 3.8, 12.5) higher among those admitted to NICU. Smaller studies were significantly associated with higher prevalences (Egger's test: $p = 0.02$). Sensitivity and specificity ranged from 89-100% and 92-100% respectively, positive predictive values from 2-84%, with all negative predictive values 100%.

LIMITATIONS: Results are generalisable to VHD countries only. Estimates and inferences were limited by available data.

CONCLUSIONS: In VHD countries, 1 per 1000 screened newborns require referral to clinical services for PCHL. Prevalence is higher in those admitted to NICU. Improved reporting would support further examination of screen performance and child demographics.

Int J Pediatr Otorhinolaryngol. 2019 Jun;121:99-108. doi: 10.1016/j.ijporl.2019.01.012. Epub 2019 Mar 8.

The prevalence of deafness-associated mutations in neonates: A meta-analysis of clinical trials.

Chen S, Liang Z, Chen B, Yao F, Huang G, Zhu J, Li Y, Gao S, Zhao Q.

OBJECTIVE: The causative genes associated with autosomal recessive non-syndromic hearing loss (ARNSHL) have been identified, in order of prevalence are GJB2, SLC26A4, MYO15A, OTOF, CDH23, and TMC1. To evaluate the prevalence of deafness-associated mutations in neonates and the clinical value of screening, we performed a meta-analysis of clinical trials.

METHODS: The main criteria used to select articles was that the studies were designed to detect deafness genetic mutations in Chinese's neonates, and the screening kits were designed to detect 9 or 20 sites in four deafness-causative genes. The combined effect of genetic screening was measured by the pooled prevalence of mutations with 95% confidence intervals (CIs). The Random Model was used to estimate the pooled prevalence of mutations.

RESULTS: We included 18 studies (a total of 261766 neonates) from studies using 9-mutation screening kit, and 15 studies (a total of 131158 neonates) from studies using the 20-mutation screening kit to conduct meta-analysis. The Random Model was used to estimate the pooled prevalence of mutations due to large heterogeneity (9 sites: $I^2 = 89.1\%$, $P = 0.0000$; 20 sites: $I^2 = 97.3\%$, $P = 0.0002$). The pooled prevalence of mutations in 9 sites group was 0.043 (95%CI:0.039-0.047, $Z = 21.49$, $P = 0.000$) and 20 sites group was 0.047(95%CI:0.041-0.053, $Z = 15.84$, $P = 0.000$).

CONCLUSIONS: The prevalence of deafness-associated mutations in neonates in China is 4.7%; Based on the current detection technology and deafness genetics knowledge, it may be more reasonable to offer 1494C>T and 1555A>G mutation screening to pregnant women. Decision makers should think about how to use the current deafness genetic screening to amplify the effectiveness of hearing screening.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109664. doi: 10.1016/j.ijporl.2019.109664. Epub 2019 Aug 30.

Referral rates for newborn hearing screening based on the test time.

Chung YS, Oh SH, Park SK.

OBJECTIVES: To suggest the optimal timing for newborn hearing screening to obtain an ideal and stable referral rate in well babies and in babies in neonatal intensive care units (NICU).

METHODS: This study analyzed nationwide hearing screening data of 71,596 newborns in the low-income families who received support from the government in 2017. The referral rate for neonatal hearing screening, based on the period from birth to screening, was compared in well babies ($n = 68,206$) who were born healthy or hospitalized in the NICU for < 5 days and moved to a well-baby nursery and babies ($n = 3,390$) who were in the NICU for ≥ 5 days.

RESULTS: In well babies, most screenings (73.1%) were performed in the first three days of life. Most babies (74%) in the NICU underwent screening within the first 20 days after birth. The referral rates for babies at 32-60 days after birth differed from those of other screening days, except for the rates at 21-31 days after birth. Referral rates for NHS at > 60 days after birth were significantly higher than those for other periods in babies overall. However, the incidence of hearing loss according to age did not differ significantly within or between groups.

CONCLUSIONS: Referral rates based on the period from birth to screening were significantly different between well babies and NICU babies. To reduce and ensure stable referral rates, we recommend that the hearing screening be performed between 2 and 20 days after birth for well babies and between 5 and 31 days after birth for NICU babies.

Newborn hearing screening at the Neonatal Intensive Care Unit and Auditory Brainstem Maturation in preterm infants.

Ciorba A, Hatzopoulos S, Corazzi V, Cogliandolo C, Aimoni C, Bianchini C, Stomeo F, Pelucchi S.

OBJECTIVES: Aim of this study is to report and discuss the results of 4 years of Newborn hearing screening (NHS) program at the Neonatal Intensive Care Unit (NICU), particularly evaluating the clinical ABR results.

METHODS: Retrospective study. NHS data from NICU newborns, admitted for ≥ 5 days, in the period from January 1st 2013 and December 31st 2016, were retrieved and analyzed. NHS results were classified as following: (i) "pass" when both ears for both the a-TEOAE (automated Transient-Evoked Otoacoustic Emissions) and the a-ABR (automated Auditory Brainstem Response) protocol resulted as "pass"; (ii) "fail" when one ear, at either one of the two performed tests resulted as "fail"; (iii) "missing" when the newborns were not tested with both protocols. All "fail" and "missing" newborns were retested (with both tests): in the case of a second "fail" result, a clinical ABR was performed within a period of 3 months.

RESULTS: A total of 1191 newborns were screened. From those, 1044/1191 resulted as "pass", 108/1191 as "fail", and 39/1191 as "missing". During the re-testing of these 147 newborns, 43 were assigned as "missing", 63 were assigned as "pass" (showing bilaterally a wave V identifiable within 30 dB nHL) and 25 failed the retest and/or did not present an identifiable wave V within 30 dB nHL. Among the 147 retested infants, we identified a group of 16 subjects who resulted as NHS "refer" and who, during the audiological follow-up, showed either: (i) a unilateral or bilateral wave V identifiable over 30 dB nHL, at the first clinical ABR assessment; or (ii) a bilateral wave V identifiable within 30 dB nHL, in a following clinical ABR test during the first year of life. These 16 subjects were defined to have an 'Auditory Brainstem Maturation' issue.

CONCLUSIONS: A possible "maturation" of the ABR response (and therefore of the auditory pathway) has been hypothesised in 16 out of 1191 infants (1.3%). A delay of the auditory pathway maturation in preterm babies compared to term newborns has already been suggested in the literature. A possible delay of the NHS retest could be considered, in selected cases, with significant savings in economic resources and parental anxiety.

Pharmacogenomics J. 2019 Oct 31. doi: 10.1038/s41397-019-0113-1. [Epub ahead of print]

Genetic variation of cisplatin-induced ototoxicity in non-cranial-irradiated pediatric patients using a candidate gene approach: The International PanCareLIFE Study.

Clemens E, Broer L, Langer T, Uitterlinden AG, de Vries ACH, van Grotel M, Pluijm SFM, Binder H, Byrne J, Broeder EVD, Crocco M, Grabow D, Kaatsch P, Kaiser M, Kenborg L, Winther JF, Rechnitzer C, Hasle H, Kepak T, van der Kooi AF, Kremer LC, Kruseova J, Kuehni CE, van der Pal H, Parfitt R, Deuster D, Matulat P, Spix C, Tillmanns A, Tissing WJE, Maier L, Am Zehnhoff-Dinnesen A, Zolk O, van den Heuvel-Eibrink MM; PanCareLIFE consortium.

Ototoxicity is a common side effect of platinum treatment and manifests as irreversible, high-frequency sensorineural hearing loss. Genetic association studies have suggested a role for SNPs in genes related to the disposition of cisplatin or deafness. In this study, 429 pediatric patients that were treated with cisplatin were genotyped for 10 candidate SNPs. Logistic regression analyses revealed that younger age at treatment (≤ 5 years vs > 15 years: OR: 9.1; 95% CI: 3.8-21.5; $P = 5.6 \times 10^{-7}$) and higher cumulative dose of cisplatin (> 450 vs ≤ 300 mg/m²: OR: 2.4; 95% CI: 1.3-4.6; $P = 0.007$) confer a significant risk of ototoxicity. Of the SNPs investigated, none of them were significantly associated with an increase of ototoxicity. In the meta-analysis, ACYP2 rs1872328 (OR: 3.94; 95% CI: 1.04-14.03; $P = 0.04$) and SLC22A2 rs316019 (OR: 1.46; 95% CI: 1.07-2.00; $P = 0.02$) were associated with ototoxicity. In order to increase the understanding of the association between SNPs and ototoxicity, we propose a polygenic model, which takes into account multiple interacting genes of the cisplatin pathway that together confer an increased risk of ototoxicity.

Vaccines (Basel). 2019 Sep 26;7(4). pii: E129. doi: 10.3390/vaccines7040129.

Impact of Maternal Immunity on Congenital Cytomegalovirus Birth Prevalence and Infant Outcomes: A Systematic Review.

Coppola T, Mangold JF, Cantrell S, Permar SR.

Congenital cytomegalovirus (cCMV) is the leading non-genetic cause of sensorineural hearing loss (SNHL), and efforts are geared towards prevention through vaccine development. Transmission rates following primary maternal infection occur at rates of 30-40%, however reported placental rates upon non-primary maternal infection is reported to be less than $< 4\%$. There is significant debate about whether this reduction in transmission rate is due to pre-existing maternal immunity, which could identify possible immunologic targets for vaccines. To address this question, we performed a systemic review of the literature using Preferred Reporting Items for Systematic Review and Analysis (PRISMA) guidelines. We identified cohort studies in high CMV seroprevalent

(>80%) areas or in developing regions that examined a cohort of at least 50 infants for congenital CMV acquisition. We identified 19 articles that met criteria and were further categorized based on pre-conception serology, maternal seroprevalence, or previously known seroprevalence. Birth prevalence rates ranged from 0.4% to 6% (median 1.1%), with the studies reporting on clinical outcome (16/19 studies) noting the majority of infected infants as asymptomatic. We also utilized a recent study that differentiated primary maternal infections from chronic infections in a highly seropositive population to calculate a placental transmission rate in women with pre-existing immunity compared to that of no pre-existing immunity. This work confirms a low cCMV birth prevalence in highly seropositive populations, indicates via a calculated placental transmission rate that the CMV placental transmission rate is lower in non-primary infection than that of primary infection, and reveals gaps in data for further research aiming to identify targets for vaccine development.

HNO. 2019 Oct;67(10):750-759. doi: 10.1007/s00106-019-00729-8.

Etiology and therapy indication for cochlear implantation in children with single-sided deafness : Retrospective analysis.

Cushing SL, Gordon KA, Sokolov M, Papaioannou V, Polonenko M, Papsin BC.

OBJECTIVE: The characteristics of children with single-sided deafness (SSD) who become candidates for unilateral cochlear implantation (uCI) were identified.

STUDY DESIGN: In all, 118 children with SSD presenting from 2013-2019 to a tertiary pediatric children's hospital were retrospectively assessed regarding candidacy for uCI.

RESULTS: Of the 118 children, 103 had completed uCI candidacy assessment, while 15 were undergoing this assessment at the time of review. More than half of children did not go on to implantation (63/103, 61%), with the 2 main reasons being (1) half (31/63) did not meet candidacy criteria for implantation, most commonly due to cochlear nerve aplasia/hypoplasia (31/82 who were assessed with MRI, 38%) and (2) families (30/103; 29%) declined participation in the surgical arm of the trial. The most common etiologies of SSD in the 37/103 (36%) children who both met candidacy and consented to implantation were congenital cytomegalovirus (cCMV; 16/37, 43%), unknown (6/37, 16%), cochleovestibular anomaly and trauma (each 5/37, 14%).

CONCLUSIONS: Many children with SSD who present for implant candidacy assessment do not ultimately receive uCI. Major factors contributing to noncandidacy are cochlear nerve aplasia and parental acceptance of the intervention. While approximately half of children with SSD in our cohort were candidates for implantation, only 1/3 of the total cohort proceeded with implantation with the main predictors of acceptability of this intervention being an etiology (i.e., cCMV) that carries risk of progressive deterioration in the better hearing ear or SSD that was sudden in onset. These findings provide important insight into this new population of cochlear implant users and the emerging acceptance of intervention in children with SSD.

Int J Audiol. 2019 Dec;58(12):834-850. doi: 10.1080/14992027.2019.1632499. Epub 2019 Jul 2.

Genetic screening as an adjunct to universal newborn hearing screening: literature review and implications for non-congenital pre-lingual hearing loss.

D'Aguillo C, Bressler S, Yan D, Mittal R, Fifer R, Blanton SH, Liu X.

OBJECTIVE: Universal newborn hearing screening (UNHS) uses otoacoustic emissions testing (OAE) and auditory brainstem response testing (ABR) to screen all newborn infants for hearing loss (HL), but may not identify infants with mild HL at birth or delayed onset HL. The purpose of this review is to examine the role of genetic screening to diagnose children with pre-lingual HL that is not detected at birth by determining the rate of children who pass UNHS but have a positive genetic screening. This includes a summary of the current UNHS and its limitations and a review of genetic mutations and screening technologies used to detect patients with an increased risk of undiagnosed pre-lingual HL.

DESIGN: Literature review of studies that compare UNHS with concurrent genetic screening.

STUDY SAMPLE: Infants and children with HL.

RESULTS: Sixteen studies were included encompassing 137,895 infants. Pathogenic mutations were detected in 8.66% of patients. In total, 545 patients passed the UNHS but had a positive genetic screening. The average percentage of patients who passed UNHS but had a positive genetic screening was 1.4%.

CONCLUSIONS: This review demonstrates the positive impact of concurrent genetic screening with UNHS to identify patients with pre-lingual HL.

Concurrent Hearing and Genetic Screening of 180,469 Neonates with Follow-up in Beijing, China.

[Dai P](#), [Huang LH](#), [Wang GJ](#), [Gao X](#), [Qu CY](#), [Chen XW](#), [Ma FR](#), [Zhang J](#), [Xing WL](#), [Xi SY](#), [Ma BR](#), [Pan Y](#), [Cheng XH](#), [Duan H](#), [Yuan YY](#), [Zhao LP](#), [Chang L](#), [Gao RZ](#), [Liu HH](#), [Zhang W](#), [Huang SS](#), [Kang DY](#), [Liang W](#), [Zhang K](#), [Jiang H](#), [Guo YL](#), [Zhou Y](#), [Zhang WX](#), [Lyu F](#), [Jin YN](#), [Zhou Z](#), [Lu HL](#), [Zhang X](#), [Liu P](#), [Ke J](#), [Hao JS](#), [Huang HM](#), [Jiang D](#), [Ni X](#), [Long M](#), [Zhang L](#), [Qiao J](#), [Morton CC](#), [Liu XZ](#), [Cheng J](#), [Han DM](#).

Concurrent hearing and genetic screening of newborns is expected to play important roles not only in early detection and diagnosis of congenital deafness, which triggers intervention, but also in predicting late-onset and progressive hearing loss and identifying individuals who are at risk of drug-induced HL. Concurrent hearing and genetic screening in the whole newborn population in Beijing was launched in January 2012. This study included 180,469 infants born in Beijing between April 2013 and March 2014, with last follow-up on February 24, 2018. Hearing screening was performed using transiently evoked otoacoustic emission (TEOAE) and automated auditory brainstem response (AABR). For genetic testing, dried blood spots were collected and nine variants in four genes, GJB2, SLC26A4, mtDNA 12S rRNA, and GJB3, were screened using a DNA microarray platform. Of the 180,469 infants, 1,915 (1.061%) were referred bilaterally or unilaterally for hearing screening; 8,136 (4.508%) were positive for genetic screening (heterozygote, homozygote, or compound heterozygote and mtDNA homoplasmy or heteroplasmy), among whom 7,896 (4.375%) passed hearing screening. Forty (0.022%) infants carried two variants in GJB2 or SLC26A4 (homozygote or compound heterozygote) and 10 of those infants passed newborn hearing screening. In total, 409 (0.227%) infants carried the mtDNA 12S rRNA variant (m.1555A>G or m.1494C>T), and 405 of them passed newborn hearing screening. In this cohort study, 25% of infants with pathogenic combinations of GJB2 or SLC26A4 variants and 99% of infants with an m.1555A>G or m.1494C>T variant passed routine newborn hearing screening, indicating that concurrent screening provides a more comprehensive approach for management of congenital deafness and prevention of ototoxicity.

Effects of Early Acoustic Hearing on Speech Perception and Language for Pediatric Cochlear Implant Recipients.

[Davidson LS](#), [Geers AE](#), [Uchanski RM](#), [Firszt JB](#).

PURPOSE: The overall goal of the current study was to identify an optimal level and duration of acoustic experience that facilitates language development for pediatric cochlear implant (CI) recipients—specifically, to determine whether there is an optimal duration of hearing aid (HA) use and unaided threshold levels that should be considered before proceeding to bilateral CIs.

METHOD: A total of 117 pediatric CI recipients (ages 5–9 years) were given speech perception and standardized tests of receptive vocabulary and language. The speech perception battery included tests of segmental perception (e.g., word recognition in quiet and noise, and vowels and consonants in quiet) and of suprasegmental perception (e.g., talker and stress discrimination, and emotion identification). Hierarchical regression analyses were used to determine the effects of speech perception on language scores, and the effects of residual hearing level (unaided pure-tone average [PTA]) and duration of HA use on speech perception.

RESULTS: A continuum of residual hearing levels and the length of HA use were represented by calculating the unaided PTA of the ear with the longest duration of HA use for each child. All children wore 2 devices: Some wore bimodal devices, while others received their 2nd CI either simultaneously or sequentially, representing a wide range of HA use (0.03–9.05 years). Regression analyses indicate that suprasegmental perception contributes unique variance to receptive language scores and that both segmental and suprasegmental skills each contribute independently to receptive vocabulary scores. Also, analyses revealed an optimal duration of HA use for each of 3 ranges of hearing loss severity (with mean PTAs of 73, 92, and 111 dB HL) that maximizes suprasegmental perception.

CONCLUSIONS: For children with the most profound losses, early bilateral CIs provide the greatest opportunity for developing good spoken language skills. For those with moderate-to-severe losses, however, a prescribed period of bimodal use may be more advantageous for developing good spoken language skills.

Screening for Language Delay between 6 Months and 3 Years of Corrected Age in Very Low Birth Weight Children.

[Debata P](#), [Kumar J](#), [Mukhopadhyay K](#).

OBJECTIVES: To screen for language delay in very low birth weight (VLBW) children between 6 months to 3 years using Language Evaluation Scale Trivandrum, 0–3 years.

METHODS: VLBW inborn neonates at a corrected age of 6 months to 3 years visiting follow-up clinic were

enrolled. Children with hearing loss were excluded. Prevalence and predictors of language delay were ascertained.

RESULTS: Of 200 enrolled subjects, out of the 1400 VLBW discharged, 64 (32%) had language delay. On multivariate analysis, late onset sepsis, patent ductus arteriosus and poor socioeconomic status were significant predictors of language delay. Abnormal neurological examination and suspect development were also associated with language delay.

CONCLUSIONS: In VLBW children, the frequency of language delay is quite high. These children should be screened for language delay.

OTO Open. 2019 May 29;3(2):2473974X19849874. doi: 10.1177/2473974X19849874. eCollection 2019 Apr-Jun.

[Congenital Cytomegalovirus and Hearing Loss: A Pilot Cross-Sectional Survey of Otolologists' and Pediatric Otolaryngologists' Knowledge.](#)

[Dedhia K](#), [Tomlinson J](#), [Murray N](#), [Park A](#).

OBJECTIVE: To evaluate pediatric otolaryngologists, neurotologists, and otologists on awareness and knowledge of congenital cytomegalovirus (cCMV).

STUDY DESIGN: Pilot cross-sectional online survey.

SETTING: Otolaryngology practices.

SUBJECTS AND METHODS: An electronic multiple-choice questionnaire was sent email listserv to physician members of the American Society of Pediatric Otolaryngology and American Otological Society. The survey assessed demographics, physician awareness, and practice patterns. Data were collected and analyzed.

RESULTS: Seventy (14.5%) pediatric otolaryngologists and otologists responded. All responded that they are familiar with cCMV. Most were familiar with symptoms associated with cCMV with the exception of petechia/purpura. Less than 50% knew the incidence/natural history of cCMV-induced hearing loss. Only 63% knew that saliva or urine polymerase chain reaction/culture should be performed prior to 3 weeks of age. Less than half knew the indications for dry blood spot testing, and many incorrectly recommended serologic saliva or urine testing in a child >3 weeks old. Most respondents do not offer any diagnostic testing for cCMV or referral for antiviral therapy for those who may benefit from this treatment. Most either did not know the cCMV screening policy or did not have one at their institution.

CONCLUSION: Despite a relatively low overall response rate, this study suggests several knowledge gaps and underutilization of cCMV testing by physicians who frequently encounter pediatric hearing loss. The findings from this pilot study demonstrate the need for further educational directives focused on cCMV to improve knowledge and incorporation of cCMV best practices.

Otolaryngol Clin North Am. 2020 Feb;53(1):103-113. doi: 10.1016/j.otc.2019.09.005. Epub 2019 Oct 21.

[Auditory Brainstem Implantation: Candidacy Evaluation, Operative Technique, and Outcomes.](#)

[Deep NL](#), [Roland JT Jr](#).

Auditory brainstem implants (ABIs) stimulate the auditory system at the cochlear nucleus, bypassing the peripheral auditory system including the auditory nerve. They are used in patients who are not cochlear implant candidates. Current criteria for use in the United States are neurofibromatosis type 2 patients 12 years or older undergoing first- or second-side vestibular schwannoma removal. However, there are other nontumor conditions in which patients may benefit from an ABI, such as bilateral cochlear nerve aplasia and severe cochlear malformation not amenable to cochlear implantation. Recent experience with ABI in the pediatric population demonstrates good safety profile and encouraging results.

Braz J Otorhinolaryngol. 2019 Jul 4. pii: S1808-8694(19)30038-2. doi: 10.1016/j.bjorl.2019.04.009. [Epub ahead of print]

[Cortical auditory evoked potential in assessment of neonates: a study about minimum level of responses in term and preterm newborns.](#)

[Didoné DD](#), [Oliveira LS](#), [Durante AS](#), [Almeida K](#), [Garcia MV](#), [Riesgo RDS](#), [Sleifer P](#).

INTRODUCTION: The study of the threshold level of cortical auditory response in adults has been investigated in previous studies. Due to maturational issues, little is known about these responses in neonates. Technological advances with automatic analysis devices now allow investigation in specific populations. Thus, new studies are needed to establish the feasibility of using this auditory potential to identify the lowest levels of responses in children.

OBJECTIVE: Verify and compare latency and amplitude in 80dBnNA and the minimum level of cortical auditory response in term and preterm neonates.

METHODS: A cross-sectional, comparative study involving 59 neonates, 35 full-term births and 24 preterm births, with positive results in the Neonatal Hearing Screening. The Hearlab system was used to investigate the P1i auditory potential with tone burst stimulus at frequencies of 500, 1000, 2000 and 4000Hz. The minimum response level search ranged from 80 to 0dBNA and was detected automatically. The results were compared between groups, evaluating the latency and amplitude in 80dBNA and the minimum level of cortical auditory response.

RESULTS: The mean values obtained for the minimum level of cortical auditory response in term group were 26 ± 8.81 ; 26.14 ± 6.97 ; 29 ± 7.65 and 29.43 ± 7.04 dBNA and for preterm neonates of 31.96 ± 10.41 ; 34.13 ± 11.34 ; 33.64 ± 11.03 and 37.73 ± 11.92 dBNA, for the frequencies of 500, 1000, 2000 and 4000Hz, respectively. There was a difference between groups for the latency of P1i at 4000Hz and the minimum response levels at 500, 1000 and 4000Hz, with higher values for preterm infants.

CONCLUSION: It was possible to obtain latency and amplitude values at 80dBnNA and the minimum level of cortical response in term and preterm newborns, with different results between groups, with higher values in those born preterm.

Ochsner J. 2019 Summer;19(2):123-130. doi: 10.31486/toj.18.0095.

[Congenital Cytomegalovirus Infection.](#)

[Dietrich ML](#), [Schieffelin JS](#).

BACKGROUND: Congenital cytomegalovirus (cCMV) is the leading cause of nongenetic congenital hearing loss in much of the world and a leading cause of neurodevelopmental disabilities. Infected babies can be born to women who are seropositive and seronegative prior to pregnancy, and the incidence is approximately 0.6%-0.7% in the United States. Symptoms vary from mild to severe, and hearing loss can be delayed in onset and progressive.

METHODS: We reviewed the literature to summarize the epidemiology, clinical manifestations, diagnosis, treatment, and future directions of cCMV.

RESULTS: The best way to diagnose the infection is with polymerase chain reaction of urine or saliva within 3 weeks after birth, followed by a repeat confirmatory test if positive. Moderately to severely symptomatic neonates should be treated for 6 months with valganciclovir, and some practitioners also choose to treat infants who have isolated hearing loss only. Treatment is not recommended for asymptomatic infants. All infected infants should be screened for hearing loss and neurodevelopmental sequelae. Universal and targeted screening may be cost effective. Currently, no vaccine is commercially available, although multiple candidates are under study.

CONCLUSION: Congenitally acquired cytomegalovirus is found in all communities around the world with a disease burden that is greater than many other well-known diseases. Advances are being made in prevention and treatment; however, improved awareness of the disease among clinicians and patients is needed.

J Multidiscip Healthc. 2019 Aug 1;12:607-620. doi: 10.2147/JMDH.S175713. eCollection 2019.

[Growth in CHARGE syndrome: optimizing care with a multidisciplinary approach.](#)

[Dijk DR](#), [Bocca G](#), [van Ravenswaaij-Arts CM](#).

CHARGE (Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and/or development, Genital hypoplasia, Ear anomalies including hearing loss) syndrome is a rare syndrome with an incidence of approximately 1:15,000 newborns. It is caused by pathogenic variants in the *CHD7* gene and clinically characterized by a wide range of anomalies with variable expression. Growth retardation affects 60-72% of children with CHARGE syndrome, making it one of the most prominent medical issues in the syndrome. Growth retardation in CHARGE syndrome is thought to be multifactorial and can be influenced by almost all co-morbidities, requiring a multidisciplinary approach to the different medical problems. In this systematic review, we describe what is currently known about growth in CHARGE syndrome and how it is influenced by commonly seen clinical problems including feeding difficulties, hypogonadotropic hypogonadism and growth hormone deficiency. Furthermore, we provide recommendations for a multidisciplinary approach.

StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2019 Oct 29.

[Pediatric Hearing Loss.](#)

[Dimitrov L](#), [Gossman WG](#).

Pediatric hearing loss is a broad category that covers a wide range of pathologies. Early detection and prompt management are essential for the development of normal language and psychosocial functioning, as well as to identify potentially reversible causes or other underlying problems. Hearing is measured in decibels, and the severity of the hearing loss is graded by hearing thresholds. The normal hearing range is 0-20 decibels (dB)

which equates to being able to perceive sound quieter than a whisper. Mild hearing loss corresponds to a range of 20-39 dB, moderate 40-69 dB, severe 70-89 dB and profound is greater than 90 dB. There are three main types of hearing loss; conductive, sensorineural and mixed. The former typically occurs due to a problem transmitting sounds at the level of the external or middle ear. The major cause of conductive hearing loss in children is otitis media with effusion (glue ear). Sensorineural hearing loss results from a disruption of the auditory pathway at any point from the cochlea of the inner ear through to the brainstem, and despite being relatively uncommon in children as a whole, it is the primary cause of permanent hearing loss in the pediatric population. Mixed hearing loss occurs when there are both conductive and sensorineural components.

Ear Nose Throat J. 2019 Sep;98(8):482-485. doi: 10.1177/0145561319840573. Epub 2019 May 29.

[Correlation Between Sensorineural Hearing Loss and Chronic Otorrhea.](#)

[Dobrianskyj FM, Dias Gonçalves ÍR, Tamaoki Y, Mitre EI, Quintanilha Ribeiro FA.](#)

Many studies have been trying to correlate chronic otorrhea, both in children and in adults, with the sensorineural hearing loss in the affected ear, but have been obtaining contradictory results. This loss might be due to the likely toxicity of the bacteria involved, effects of inflammatory cytokines, or constant use of ototoxic antibiotics. All the studies evaluated up to the present date compared the affected ear with the normal contralateral ear. From the digitized archive of otological surgery files of the Department of Otorhinolaryngology, the ears of patients with chronic otorrhea were evaluated visually and compared with the normal contralateral ears. Ears with otorrhea were also compared to ears with dry tympanic perforation of other patients. Ears with suppuration were evaluated for cholesteatoma. The duration of otorrhea was taken into account. The sensorineural hearing threshold was evaluated for the frequencies of 500, 1000, 2000, and 4000 Hz. A total of 98 patients with chronic otorrhea and 60 with dry tympanic membrane perforation were evaluated. From a statistical study, a correlation between sensorineural hearing loss and the chronic otorrhea was observed, in comparison both with contralateral normal ears and with dry perforated ears of other patients. There was no relationship with the duration of suppuration or with whether this was due to cholesteatoma. Sensorineural hearing loss occurs in ears with chronic otorrhea. The duration of otorrhea and the etiology of suppuration did not influence the hearing loss. The great majority of otorrhea cases begin during childhood.

Braz J Otorhinolaryngol. 2019 Sep 17. pii: S1808-8694(19)30095-3. doi: 10.1016/j.bjorl.2019.08.001. [Epub ahead of print]

[Otoacoustic emissions in neonates exposed to smoke during pregnancy.](#)

[Durante AS, Nascimento CMD, Lopes C.](#)

INTRODUCTION: The toxic substances present in cigarette smoke can damage cochlea hair cells. This effect has been investigated by measuring otoacoustic emissions.

OBJECTIVE: To investigate the impact of stimuli on otoacoustic emissions, comparing neonates with and without exposure to cigarette smoke during pregnancy.

METHODS: Transient-evoked otoacoustic emissions, evoked by a click stimulus, and distortion product otoacoustic emissions, evoked by two tones tests were conducted in both ears, using an Interacoustic TITAN device. The study included 105 neonates divided into two groups: a study group, comprising 47 neonates exposed to smoke during pregnancy; and a control group comprised of 58 neonates who were not exposed. All participants had normal neonatal hearing screening.

RESULTS: No statistical differences in distortion product otoacoustic emissions response levels were found between the groups. In the transient-evoked otoacoustic emissions tests lower response levels were observed in the study group than the control group in frequency band analysis of the right ear, with statistically significant differences in signals and signal-noise ratio (except at 1 kHz).

CONCLUSION: The impact of smoking exposure could be analyzed through transient-evoked otoacoustic emissions in neonates. The group effect of smoke exposure during pregnancy was evidenced by a reduction in transient-evoked otoacoustic emissions levels. This same effect was not observed for the analyses performed on distortion product otoacoustic emissions levels.

Front Psychol. 2019 Sep 10;10:2083. doi: 10.3389/fpsyg.2019.02083. eCollection 2019.

[Effects of a Manual Response Requirement on Early and Late Correlates of Auditory Awareness.](#)

[Eklund R, Gerdfeldt B, Wiens S.](#)

In hearing, two neural correlates of awareness are the auditory awareness negativity (AAN) and the late positivity (LP). These correlates of auditory awareness are typically observed with tasks in which subjects are required to report their awareness with manual responses. Thus, the correlates may be confounded by this manual response requirement. We manipulated the response requirement in a tone detection task ($N = 52$). Tones were

presented at each subject's individual awareness threshold while high-density electroencephalography (EEG) activity was recorded. In one response condition, subjects pushed a button if they were aware of the tone and withheld responding if they were unaware of the tone. In the other condition, subjects pushed a button if they were unaware of the tone and withheld responding if they were aware of the tone. To capture AAN and LP, difference waves were computed between aware and unaware trials, separately for trials in which responses were required and trials in which responses were not required. Results suggest that AAN and LP are unaffected by the response requirement. These findings imply that in hearing, early and late correlates of awareness are not confounded by a manual response requirement. Furthermore, the results suggest that AAN originates from bilateral auditory cortices, supporting the view that AAN is a neural correlate of localized recurrent processing in early sensory areas.

Otolaryngol Head Neck Surg. 2019 Oct;161(4):672-682. doi: 10.1177/0194599819849917. Epub 2019 Jun 18.

Expanding Access: Cost-effectiveness of Cochlear Implantation and Deaf Education in Asia.

Emmett SD, Sudoko CK, Tucci DL, Gong W, Saunders JE; Global HEAR (Hearing Loss Evaluation, Advocacy, and Research) Collaborative; Akhtar N, Bhutta MF, Touch S, Pradhananga RB, Mukhtar N, Martinez N, Martinez FD, Ramos H, Kameswaran M, Kumar RNS, Soekin S, Prepageran N.

OBJECTIVE: To determine the cost-effectiveness of cochlear implantation (CI) with mainstream education and deaf education with sign language for treatment of children with profound sensorineural hearing loss in low- and lower-middle income countries in Asia.

STUDY DESIGN: Cost-effectiveness analysis.

SETTING: Bangladesh, Cambodia, India, Indonesia, Nepal, Pakistan, Philippines, and Sri Lanka participated in the study.

SUBJECTS AND METHODS: Costs were obtained from experts in each country with known costs and published data, with estimation when necessary. A disability-adjusted life-years model was applied with 3% discounting and 10-year length of analysis. A sensitivity analysis was performed to evaluate the effect of device cost, professional salaries, annual number of implants, and probability of device failure. Cost-effectiveness was determined with the World Health Organization standard of cost-effectiveness ratio per gross domestic product (CER/GDP) per capita <3.

RESULTS: Deaf education was cost-effective in all countries except Nepal (CER/GDP, 3.59). CI was cost-effective in all countries except Nepal (CER/GDP, 6.38) and Pakistan (CER/GDP, 3.14)-the latter of which reached borderline cost-effectiveness in the sensitivity analysis (minimum, maximum: 2.94, 3.39).

CONCLUSION: Deaf education and CI are largely cost-effective in participating Asian countries. Variation in CI maintenance and education-related costs may contribute to the range of cost-effectiveness ratios observed in this study.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109647. doi: 10.1016/j.ijporl.2019.109647. Epub 2019 Aug 21.

Early detection of neonatal hearing loss by otoacoustic emissions and auditory brainstem response over 10 years of experience.

Escobar-Ipuz FA, Soria-Bretones C, García-Jiménez MA, Cueto EM, Torres Aranda AM, Sotos JM.

OBJECTIVES: A number of different screening protocols for detecting neonatal hearing loss currently exist. We present our 10 years of experience with using auditory brainstem response (ABR) complementary to otoacoustic emissions (OAEs) in the three phases hearing screening process in our hospital. Furthermore, we want to demonstrate the usefulness of these screening techniques used in combination, that remain valid to identify cases of neonatal hearing loss and meet the well-established program quality criteria for these screening protocols.

METHODS: Data were collected retrospectively from patient record forms completed on 9698 newborns from 2007 to 2017. The screening protocol for neonatal hearing loss in our centre is carried out in three phases. First phase, prior to discharge from the hospital, consists of carrying out the OAE evaluation on the newborn. Second phase is carried out in the paediatric consultation department. There, the newborns who did not pass the first phase are again studied with OAE. If this phase is not passed either, the child is referred to a third phase for the realization of ABR, in the clinical neurophysiology service. Newborns with risk factors for hearing loss, identified in the first phase, also go on to this third phase. When this hearing threshold exceeds 30 dB, it is considered abnormal. Cases with abnormal ABR, has a re-test conducted within the next six months from the initial ABR assessment.

RESULTS: A total of 9390 (97.1%) OAEs were performed during first phase, with 8245 newborns (87.8%) passing the screening test, while 1145 children (12.1%) presented an abnormal OAE and were included in the second screening phase. Second phase involving a repeat OAE examination performed on 1077 newborns (94%). In this second phase, 941 newborns (87.3%) passed the test. Nevertheless, 136 newborns (12.6%) failing the retest and were referred to continue on to phase three. Furthermore, 181 newborns (1.8%) presented high-

risk factors at birth and were also included in this third phase. However, in the registries of children referred to this phase, only 255 (80%) ABR evaluations were confirmed. In total, 227 newborns (2.3%) were missed from the first to third phases of the screening process. According to the database of the clinical neurophysiology service, ABRs evaluations were performed in 352 newborns referred between December 2007 and December 2017. Of this sample, 38.9% were boys and 61.1% were girls. From among cases underwent ABR, 34% of newborns did not pass the OAEs. The most common risk factor was prematurity (with admission to the neonatal intensive care unit for more than five days), affecting 28%. Abnormal ABRs waveforms were found in 43.9%, with 12.3% having a sensorineural hearing loss, 26.5% showing mixed hearing loss and, conductive hearing loss being present in 61.9%. Considering sensorineural hearing loss and other types of severe hearing loss, affected patients constituted only 1.7% of the total number of individuals studied. Finally, regarding quality control of the program participation in the first phase of care included 97.2% of all newborns, yielding a third phase referral rate of 2.9%, confirmation of a diagnosis before the fourth month of life in more than 90% of cases with an average of 3.4 months of age, and a hearing impairment detection rate as an outcome indicator of 4.5%.

CONCLUSIONS: Our data are similar to those of previous studies on screening for hearing loss in newborns. We have demonstrated the advantages of carrying out this protocol in three phases using the otoacoustic emissions together with auditory brainstem response, diagnostic tools that remain as a Gold Standard. Also, we want to highlight and demonstrate the importance of interdisciplinary coordination between the paediatric and clinical neurophysiology services in the implementation of this screening protocol. The foregoing has allowed us to comply with the proposed quality indicators, reaching coverage percentages of more than 95%, confirming the diagnosis of hearing loss within the first six months of life and making timely referrals to benefit the newborns with hearing impairment by way of treatment and follow-up in the early stages of development, avoiding future disabilities.

J Trop Pediatr. 2019 Aug 1;65(4):328-335. doi: 10.1093/tropej/fmy055.

[Zika Virus Infection during Pregnancy and Sensorineural Hearing Loss among Children at 3 and 24 Months Post-Partum.](#)

[Fandiño-Cárdenas M](#), [Idrovo AJ](#), [Velandia R](#), [Molina-Franky J](#), [Alvarado-Socarras JL](#).

OBJECTIVE: The objectives of this study were to describe the findings of the auditory screening in children of mothers with ZIKV during pregnancy or suspicious of congenital ZIKV, and to determine whether hearing loss was in the first 2 years in life, regardless of whether microcephaly was also present.

METHODS: This is a cases report. The information was collected and recorded in a database between January 2016 and April 2018. We perform two auditory tests to 3 and 24 months of life. The study was developed in Aguachica (Cesar, Colombia). It is considered a high-risk area for ZIKV infection. Participants included children of mothers with confirmed ZIKV during pregnancy or suspicious of congenital ZIKV exposure of ZIKV infection during an epidemic period in a tropical area. We defined a positive case according to the epidemiological definition and clinical criteria based on maternal symptoms. However, other children of mothers without clinical signs of Zika were evaluated at the same time. The main outcome was the presence of sensorineural hearing loss.

RESULTS: The median age in the study group (n=43) was 3.5 months (rank: 0-6) and the comparison group (n: 23, children of mothers without clinical signs of ZIKV) was 3 months (rank: 0-12). Screening hearing test was done using distortion product otoacoustic emissions. At 3 months follow-up, children were evaluated using distortion product otoacoustic emissions and automatized auditory brainstem response. None of the patients evaluated in this study were found to have sensorineural hearing loss.

CONCLUSIONS: We did not find hearing loss during the first 2 years in the children whose mother showed Zika during pregnancy. We recommend these children must be assessed to closed because there is a high risk the hearing loss as it usually may occur with CMV.

Clin Infect Dis. 2019 Oct 15;69(9):1526-1532. doi: 10.1093/cid/ciy1128.

[Sequelae of Congenital Cytomegalovirus Following Maternal Primary Infections Are Limited to Those Acquired in the First Trimester of Pregnancy.](#)

[Faure-Bardon V](#), [Magny JF](#), [Parodi M](#), [Couderc S](#), [Garcia P](#), [Maillotte AM](#), [Benard M](#), [Pinquier D](#), [Astruc D](#), [Patural H](#), [Pladys P](#), [Parat S](#), [Guillois B](#), [Garenne A](#), [Bussi res L](#), [Guilleminot T](#), [Stirnemann J](#), [Ghout I](#), [Ville Y](#), [Leruez-Ville M](#).

BACKGROUND: The known relationship between the gestational age at maternal primary infection the outcome of congenital CMV is based on small, retrospective studies conducted between 1980 and 2011. They reported that 32% and 15% of cases had sequelae following a maternal primary infection in the first and second or the third trimester, respectively. We aimed to revisit this relationship prospectively between 2011 and 2017, using accurate virological tools.

METHODS: We collected data on women with a primary infection and an infected child aged at least 1 year

at the time of analysis. An accurate determination of the timing of the primary infection was based upon serial measurements of immunoglobulin (Ig) M and IgG and on IgG avidity in sera collected at each trimester. The case outcome was assessed according to a structured follow-up between birth and 48 months.

RESULTS: We included 255 women and their 260 fetuses/neonates. The dating of the maternal infection was prospective in 86% of cases and retrospective in 14%. At a median follow-up of 24 months, the proportion of sensorineural hearing loss and/or neurologic sequelae were 32.4% (95% confidence interval [CI] 23.72-42.09) after a maternal primary infection in the first trimester, 0 (95% CI 0-6.49) after an infection in the second trimester, and 0 (95% CI 0-11.95) after an infection in the third trimester ($P < .0001$).

CONCLUSIONS: These results suggest that a cytomegalovirus infection can be severe only when the virus hits the fetus in the embryonic or early fetal period. Recent guidelines recommend auditory follow-ups for at least 5 years for all infected children. This raises parental anxiety and generates significant costs. We suggest that auditory and specialized neurologic follow-ups may be recommended only in cases of a maternal infection in the first trimester.

Int J Pediatr Otorhinolaryngol. 2019 Oct 15;129:109729. doi: 10.1016/j.ijporl.2019.109729. [Epub ahead of print]

Audiological benefit and subjective satisfaction of children with the ADHEAR audio processor and adhesive adapter.

Favoreel A, Heuninck E, Mansbach AL.

OBJECTIVES: The main objective of this study was to evaluate the audiological benefit of the ADHEAR system in a group of children with a uni- or bilateral conductive hearing loss (CHL) during a short-term exposure of three weeks, and to compare it to a conventional bone conduction hearing aid (BCHA) on a softband. The secondary aim was to assess the quality of life improvement and patient satisfaction with the ADHEAR system.

METHODS AND MATERIALS: The study was designed as a prospective study with repeated measures, where each subject served as his/her own control. Ten children (4-17 y/o) with a uni- or bilateral congenital or acquired CHL were included in this study. Pure tone audiometry and speech audiometry in quiet, both unaided and aided, were performed initially with the ADHEAR system and a BCHA on a softband, and after three weeks with the ADHEAR alone. Furthermore, patient satisfaction and quality of life were assessed using the SSQ12 and the ADHEAR questionnaire.

RESULTS: The mean unaided free field hearing threshold of 50 dB HL (with 95%CI between 41.7 and 57.5 dB HL) expressed in 'Bureau International d'Audiophonologie' (BIAP), improved significantly by 22 dB (13.0-29.9) with the ADHEAR and by 23 dB (13.6-32.9) with the BCHA ($p < 0.001$). Furthermore, the mean unaided speech recognition threshold (SRT) in quiet improved significantly by 19 dB (10.3-28.1) with the ADHEAR and by 21 dB (12.6-29.4) with the BCHA ($p < 0.001$). For both audiological tests, there were no significant differences between the ADHEAR and the BCHA. After three weeks of use, the mean pure tone threshold of 28 dB HL (20.0-36.5) and the mean SRT of 47 dB SPL (41.9-51.5) with the ADHEAR system were comparable and not significantly different than the outcomes during the first visit. Speech understanding in noise and in multiple streams, sound localization and sound quality were rated significantly better with the ADHEAR, compared to the ratings without the ADHEAR system ($p < 0.001$). None of the children reported skin irritations or pain.

CONCLUSIONS: The children included in our study had significantly improved hearing thresholds, speech perception in quiet and quality of life with the ADHEAR. The device can be an effective treatment method and a valuable alternative to other BCHA for children with a CHL, although the subjective experience of each child has to be taken into account.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109684. doi: 10.1016/j.ijporl.2019.109684. Epub 2019 Sep 15.

Evaluating butterfly inlay tympanoplasty.

Ference RS, Leonard JA, Weinstock MS, Bent JP.

OBJECTIVE: To compare closure rate, reduction in air-bone-gap, and operative time of butterfly tympanoplasty (BT) to underlay tympanoplasty (UT).

METHODS: Retrospective cohort study of children (age < 18 y) undergoing Type I tympanoplasty between 2009 and 2017. Patients were excluded if they had < 6 months of follow up, mastoidectomy, fat graft or cholesteatoma.

RESULTS: Twenty-one patients (mean age 13.4) underwent BT while forty-one patients (mean age 13.5) underwent UT. The mean size of perforation in 30.6% in BT patients and 43.6% in UT patients ($p = 0.01$). Preoperative audiogram showed a similar air-bone-gap between the two groups of 31.7, 22.7, and 17.9 dB in BT vs 29.6, 24.8, and 17.6 dB in UT at 500, 1000, and 2000 Hz, respectively ($p = 0.65, 0.63, \text{ and } 0.94$). Operative time was reduced in BT as compared to UT (94.0 min vs. 150.9, $p = 0.01$). Closure rate was similar at 85.7% in BT vs 75.6% in UT patients ($p = 0.40$). Average reductions in air-bone gap were similar with 19.2, 11.7, and 13.2 dB for BT vs 16.6, 12.1, and 10.3 dB for UT at 500, 1000, and 2000 hz, respectively ($p = 0.66, 0.93, 0.40$).

CONCLUSION: BT has become a reliable tool for the pediatric otolaryngologist. This retrospective study shows that pediatric BT results in similar outcomes with reduced operative time.

Int J Pediatr Otorhinolaryngol. 2019 Sep;124:193-199. doi: 10.1016/j.ijporl.2019.05.024. Epub 2019 May 21.

Who misses the newborn hearing screening? Five years' experience in Friuli-Venezia Giulia Region (Italy).

[Feresin A](#), [Ghiselli S](#), [Marchi R](#), [Staffa P](#), [Monasta L](#), [Orzan E](#).

INTRODUCTION: Permanent hearing impairment is the most common sensory disorder in newborns. The Universal Newborn Hearing Screening (UNHS) is widely adopted as a cost-effective procedure to achieve early identification and treatment of congenital hearing impairment, with the final goal of an improved linguistic and cognitive outcome for hearing impaired children. The Italian Ministry of Health has recently comprised UNHS in the Essential Level of Health Assistance. Nevertheless, programs still vary both across and within Italian Regions in terms of coverage, testing, referral and tracking protocols. In Friuli-Venezia Giulia region the program for the early identification of newborn and childhood hearing impairment is operative since 2012. In order to minimize the lost to follow-up cases, UNHS and childhood hearing surveillance activities have been organized in close collaboration among birth centres, paediatric audiology services, territorial Family Paediatricians and the sole regional centre for paediatric hearing loss management.

MATERIAL AND METHODS: We performed a five years' retrospective analysis of the UNHS experience in Friuli-Venezia Giulia comparing the UNHS activity of year 2013 and year 2017. The focus of the study concerns the "missing" cases. Three different typologies of "miss" cases ("documentation-miss", "access-miss" and "pathway-miss") have been defined in correspondence with main reasons for their occurrence.

RESULTS: Births in Friuli-Venezia Giulia were 9465 and 8432, respectively in 2013 and 2017. International quality indicators improved with a gain of efficiency in 5 years' experience. However, "missing" cases were 486 in 2013 and 321 in 2017, mainly due to the lack of an efficient documentation system.

CONCLUSION: UNHS programs have proven to be valuable and cost-effective in Friuli-Venezia Giulia and other Italian regions. New resources and efforts are required to achieve a complete standardization and informatisation of the UNHS data to avoid documentation gaps. A possible strategy would point to the opportunity to unify data management systems for all the ongoing newborn screening programs (metabolic, hearing and visual), linking the integrated IT system with the regional repository of current datasets.

Ear Hear. 2019 May/Jun;40(3):651-658. doi: 10.1097/AUD.0000000000000647.

Early Hearing Detection and Intervention: Timely Diagnosis, Timely Management.

[Findlen UM](#), [Hounam GM](#), [Alexy E](#), [Adunka OF](#).

OBJECTIVE: A quality improvement study was completed to assess the impact of three clinical practice changes on the timing of diagnosis and intervention for congenital hearing loss.

DESIGN: A retrospective chart review was conducted for 800 infants evaluated for congenital hearing loss before and after implementing three clinical practice changes: the use of Kalman-weighted signal averaging for auditory brainstem response testing, a tone burst-prioritized testing protocol, and expediting scheduling of initial assessment. The impact of middle ear involvement on age at diagnosis and history of neonatal intensive care unit stay on age at treatment was also examined.

RESULTS: The use of Kalman-weighted signal averaging for auditory brainstem response testing, a tone burst-prioritized testing protocol, and expedited scheduling of initial assessment each resulted in a decrease of age at diagnosis. Ultimately, the age at initial assessment was the only significant predictor related to decreased timeline for diagnosis. Middle ear pathology significantly increased age at diagnosis, while history of time in the neonatal intensive care unit significantly increased the age at provision of amplification as a treatment for permanent hearing loss.

CONCLUSIONS: The technology used for assessment, clinical protocol, and timing of assessment of infants can impact the timeline for diagnosis and treatment of congenital hearing impairment. Given the significant sequelae of delayed or missed diagnosis of hearing loss in infancy, implementing clinical practice changes should be considered at pediatric diagnostic centers.

Ear Hear. 2019 May/Jun;40(3):671-679. doi: 10.1097/AUD.0000000000000651.

Service Preferences of Parents of Children With Mild Bilateral or Unilateral Hearing Loss: A Conjoint Analysis Study.

[Fitzpatrick EM](#), [Coyle D](#), [Gaboury I](#), [Durieux-Smith A](#), [Whittingham J](#), [Grandpierre V](#), [Na E](#), [Salamatmanesh M](#).

OBJECTIVE: Universal newborn hearing screening results in substantially more children with mild bilateral and unilateral hearing loss identified in the early years of life. While intervention services for children with moderate loss and greater are generally well-established, considerable uncertainty and variation surrounds the need for intervention services for children with milder losses. This study was undertaken with parents of young children

with permanent mild bilateral and unilateral hearing loss to examine their preferences for characteristics associated with intervention services.

DESIGN: Conjoint analysis, a preference-based technique, was employed to study parents' strength of preferences. Using a cross-sectional survey that consisted of eight hypothetical clinic scenarios, we invited parents to make a discrete choice (to select one of two or more different services) between available services with different characteristics. The survey was informed by qualitative interviews conducted for this purpose. The questionnaire was administered to parents receiving intervention services in the province of Ontario, Canada, who were enrolled in a mixed-methods longitudinal study examining outcomes in early-identified children with mild bilateral/unilateral hearing loss. Data were analyzed using a generalized linear model (probit link) to identify attributes of interest for the respondents. Characteristics of the children were entered into the model to control for differences in age of diagnosis, sex, laterality of hearing loss, and hearing aid use.

RESULTS: A total of 51 of 62 invited parents completed the questionnaire. All four attributes of care that were included in the survey were found to be statistically significant, that is, parents valued support for amplification, support for speech-language development, emotional support, and communication from professionals. Analysis showed greater preference for enhanced levels relating to support for speech-language development than for support for amplification. Preference for attributes relating to emotional support and communication were also greater than for support for amplification use.

CONCLUSIONS: Conjoint analysis was used to quantify parents' preferences for service attributes. Parents' values provide insights into the aspects of a service model that should receive consideration in the development of intervention programs for young children with mild bilateral or unilateral hearing loss and their families. Although parents of young children with mild bilateral or unilateral hearing loss valued several components of care, they indicated a clear preference for speech-language support compared with support for amplification use.

Pediatrics. 2019 Aug;144(2). pii: e20183095. doi: 10.1542/peds.2018-3095. Epub 2019 Jul 2.

[Hearing Loss With Congenital Cytomegalovirus Infection.](#)

[Foulon I, De Brucker Y, Buyl R, Lichtert E, Verbruggen K, Piérard D, Camfferman FA, Gucciardo L, Gordts F.](#)

OBJECTIVE: In this study, we determined the prevalence of hearing loss in 157 children with proven congenital cytomegalovirus (cCMV) infection. We looked at possible risk determinants for developing hearing loss and proposed recommendations for screening and follow-up in the newborn.

METHODS: In a prospective 22-year study, 157 children with proven cCMV infection were evaluated for sensorineural hearing loss (SNHL). The development of SNHL was correlated with the type of maternal infection (primary versus nonprimary), the gestational age of maternal primary infection, imaging findings at birth, and the presence of symptomatic or asymptomatic infection in the newborn.

RESULTS: Of all children, 12.7% had SNHL, and 5.7% needed hearing amplification because of SNHL. Improvement, progression, and fluctuations of hearing thresholds were seen in 45%, 53.8%, and 5.7% of the children, respectively. Hearing loss was more common in the case of a symptomatic infection at birth ($P = .017$), after a maternal primary infection in the first trimester of pregnancy ($P = .029$), and in the presence of abnormalities on a neonatal brain ultrasound and/or MRI ($P < .001$).

CONCLUSION: SNHL is a common sequela in children with cCMV infection. Risk factors for SNHL were primary maternal infections before the 14th week of pregnancy, the presence of a disseminated infection at birth, and imaging abnormalities in the newborn. These children may benefit from a more thorough investigation for SNHL than children who do not present with those risk factors.

Lancet Child Adolesc Health. 2019 Aug;3(8):578-584. doi: 10.1016/S2352-4642(19)30115-4. Epub 2019 May 31.

[Interventions for cisplatin-induced hearing loss in children and adolescents with cancer.](#)

[Freyer DR, Brock P, Knight K, Reaman G, Cabral S, Robinson PD, Sung L.](#)

The identification of preventive interventions that are safe and effective for cisplatin-induced ototoxicity is important, especially in children because hearing loss can impair speech-language acquisition development. Previous randomised trials assessed systemic drugs such as amifostine, sodium diethyldithiocarbamate or disulfiram, and sodium thiosulfate. Amifostine, sodium diethyldithiocarbamate, and disulfiram did not show hearing preservation. Paediatric trials assessing sodium thiosulfate showed efficacy in terms of hearing protection. The SIOPEL 6 trial consisted solely of patients with localised hepatoblastoma and no effects on survival were shown. In the ACCL0431 trial, which included heterogeneous patients, a post-hoc analysis showed significantly worse overall survival among patients who had disseminated disease receiving sodium thiosulfate than among controls, but not among those with localised disease. Intratympanically administered drugs have mainly been assessed in adults and include N-acetylcysteine and dexamethasone. Inconsistent effects of these drugs were identified but these studies were limited by design, small sample size, and statistical approach. Future studies of systemic drugs will need to consider the measurement of disease outcomes through study design and sample size, and ototoxicity endpoints should be harmonised to enhance comparability between trials.

Hearing loss in very preterm infants: should we wait or treat?

[Frezza S](#), [Catenazzi P](#), [Gallus R](#), [Gallini F](#), [Fioretti M](#), [Anzivino R](#), [Corsello M](#), [Cota F](#), [Vento G](#), [Conti G](#).

This study investigated hearing threshold changes during the first year of corrected age (CA) in infants admitted in a neonatal intensive care unit (NICU). In 5 years, 239 infants with birth weight (BW) $\leq 1,000$ gm and/or gestational age (GA) ≤ 30 weeks were enrolled. Hearing was evaluated by oto-acoustic emission (OAEs) before discharge and auditory brainstem response (ABR) within 3 months of CA. Infants affected by unilateral or bilateral hearing loss were addressed to audiological follow-up until definitive diagnosis (within 6 months of CA). Changes in hearing threshold were also carefully analysed. 207 (86.6%) infants had normal hearing while 32 infants (13.4%) showed hearing loss (HL) at the confirmative ABR evaluation (9 mild, 16 moderate, 4 severe, 3 profound). The latter showed lower GA (27.7 ± 2 vs 28.4 ± 1.2 ; $p = 0.0061$) and BW (950 ± 390 vs $1,119 \pm 326$ gm; $p = 0.0085$). At final evaluation, 15 infants (47%) recovered a normal hearing. HL was confirmed in 17 patients. Among these, 3 infants were addressed to audiological follow-up (one case of mild unilateral hearing loss (UHL) and two with moderate UHL), while in 14 cases (44%) with bilateral sensory neural hearing loss (SNHL) (7 moderate, 4 severe, 3 profound) hearing aids were prescribed. They showed significantly lower GA and longer hospital stay in the NICU in comparison with infants without indication for audiological habilitation (18 infants) (GA 26.2 ± 2.2 weeks vs 28.4 ± 2.4 ; $p = 0.01$; NICU stay 132 ± 67 vs 59 ± 7 ; $p = 0.0002$). Definitive diagnosis was obtained at 5.9 ± 1.3 months of CA. Our study confirms the importance of audiological surveillance in preterm newborns. Hearing thresholds of preterm infants with hearing loss can change during the first year of CA and we observed normalisation in 47% of our patients. Most vulnerable to permanent SNHL were very preterm infants with a longer NICU stay, while a shorter stay represents a favourable prognostic factor for hearing improvement.

Otolaryngol Pol. 2019 May 22;73(4):1-7. doi: 10.5604/01.3001.0013.1924.

Follow-up on the diagnostic level of children covered by the Universal Neonatal Hearing Screening Program in Poland, divided into voivodships.

[Greczka G](#), [Zych M](#), [Dąbrowski P](#), [Gasińska A](#), [Król A](#), [Szyfter-Harris J](#), [Szyfter W](#).

INTRODUCTION: Thanks to the Polish Universal Neonatal Hearing Screening Program (PUNHSP), all newborns in Poland undergo a free, screening hearing examination. Between 2006 and 2015, the average number of tested children per year was 373,477. According to the analysis of The Central Database (CDB), only 55.8% of the children attended the detailed hearing examinations at the second level of the Program.

AIM: The aim of this study is to analyse the dates concerning the attendance of the children at the diagnostic level of PUNHSP in different regions of Poland.

MATERIALS AND METHODS: To conduct an analysis of this fact and find out the reasons for low attendance at the second level in 2015, a telephone survey questionnaire was developed for parents who had not registered their babies for further consultation - 3,239 randomly selected parents.

RESULTS: The analysis revealed that the number of children examined at the second diagnostic level of the program is in fact much higher than the results of The Central Database show. The actual number is 83.6% as opposed to 55.8%. As a result of the telephone questionnaire some inaccuracies in the input data to the CDB were detected. The main errors in gathering the information for the CDB were incorrect OAE test result and no examination performed. Conclusion: In Poland the worst results (i.e. questionnaire results compared to CDB) for the attendance at the diagnostic level were shown in Pomorskie, Lubelskie, Mazowieckie and Podlaskie regions. In many cases there was a large discrepancy between the reality and the information in the CDB. The improvement of clarity concerning the CDB application is important in order to minimise the possibility of malformation in the CDB.

Isr Med Assoc J. 2019 Sep;21(9):607-611.

Hearing Loss and Ophthalmic Pathology in Children Diagnosed Before and after the Implementation of a Universal Hearing Screening Program.

[Gruber M](#), [Brown C](#), [Mahadevan M](#), [Neeff M](#).

BACKGROUND: Ophthalmic pathologies may further complicate the sensory input of patients with congenital hearing loss; however, data on children with coexisting impairment of vision and hearing is outdated, from before universal implementation of hearing screening programs.

OBJECTIVES: To examine the different ophthalmic pathologies among children with congenital sensorineural hearing loss (SNHL) before or after the introduction of a universal newborn hearing screening program (UNHSP).

METHODS: Retrospective cohort study was conducted of 91 children diagnosed with congenital SNHL between 2005 and 2016 in a tertiary pediatric hospital. All patients completed an ophthalmologic examination, including

assessment of visual acuity, refraction, ocular motility, slit lamp examination, and indirect funduscopy. Radiological assessment and genetic analysis were offered to all caregivers.

RESULTS: Average age at diagnosis was 4.1 years. Nineteen children (21%) were diagnosed with an ophthalmic condition, of which the most common were refractive pathologies. Diagnosis of an ophthalmic pathology was twice as likely in the pre-UNHSP era (14 children, 27%) compared to the post-UNHSP era (5 children, 13%). Out of 91 children, 57 (63%) underwent a computed tomography scan and/or magnetic resonance imaging. Imaging was positive for structural abnormalities in 23 children (40%). There was no correlation between imaging and ophthalmic conditions. Genetic analysis was performed in 67 patients (74%).

CONCLUSIONS: The ophthalmic assessment of babies and children with congenital SNHL may yield in significant numbers of children with concomitant ophthalmic pathologies. Implementation of a UNHSP allows early diagnosis and treatment of coexisting ophthalmic and hearing conditions.

Orv Hetil. 2019 Nov;160(47):1850-1855. doi: 10.1556/650.2019.31604.

[\[Verification results of objective newborn hearing screening\].](#)

[Gáborján A](#), [Götze J](#), [Küstel M](#), [Kecskeméti N](#), [Baranyi I](#), [Csontos F](#), [Tamás L](#).

Sufficient hearing is the cornerstone of the development of children's complex sensory perception, sound recognition, speech development and optimal communication skills. Hearing screening of newborns is necessary to detect congenital hearing disorders. Compulsory objective hearing screening in Hungary is a significant improvement in early diagnosis. The Audiological Department of Otorhinolaryngology, Head and Neck Surgery Clinic at Semmelweis University serves as a verification center for children identified *via* the compulsory objective newborn hearing screening and necessitates more detailed assessment. The goal of this study was to summarize the verification results of the year 2018. Case history, ENT examination, electric response measurement, impedance tests, otoacoustic emission measurement, surdopedagogical examination, and genetic examination are the basics of the diagnosis and the therapy as well. Altogether 261 newborns were examined in 2018 and 164 were subjected to audiological tests during the analyzed seven-month period. Normal hearing was detected in both ears in 77% of the cases, while hearing loss has been verified in 37 patients (23% of cases). Permanent hearing loss has been diagnosed in 19 cases, 4 unilateral and 15 bilateral. Hearing loss of sensorineural origin was confirmed in 17, conductive in 2 children. Temporary hearing loss caused by otitis media with effusion was found in further 18 children which healed spontaneously in most cases. Organized neonatal objective hearing examination has been established in Hungary in 2015. As a result, we can diagnose and provide care for children with hearing loss at the earliest stage. The National Newborn Hearing Screening Registry ensures a well-coordinated and smooth process.

Otol Neurotol. 2019 Sep;40(8):e769-e773. doi: 10.1097/MAO.0000000000002314.

[Cochlear Implant Outcomes in Large Vestibular Aqueduct Syndrome-Should We Provide Cochlear Implants Earlier?](#)

[Hall AC](#), [Kenway B](#), [Sanli H](#), [Birman CS](#).

OBJECTIVE: Examine postoperative speech perception outcomes in a large vestibular aqueduct syndrome (LVAS) patients at a major cochlear implantation center.

STUDY DESIGN: Retrospective analysis of the Sydney Cochlear Implant Centre (SCIC) database and medical records from January 1994 to December 2015 was performed.

SETTING: Tertiary referral center.

PATIENTS: Patients with a diagnosis of LVAS who received a cochlear implant (CI). Only those with speech perception outcomes recorded at least 12 months post implant were included in our analysis.

INTERVENTION(S): Therapeutic.

MAIN OUTCOME MEASURE(S): Postoperative speech perception scores.

RESULTS: Between 1994 and 2015, 176 adult and pediatric patients with a diagnosis of LVAS underwent cochlear implantation at SCIC. Postoperative Bamford-Kowal Bench (BKB) sentence test scores were obtained for 97 patients. The postoperative median BKB score was 93% with a lower quartile score of 85% and an upper quartile score of 98%. Smaller numbers were available for post-CI City University of New York (CUNY) and Consonant-Nucleus-Consonant (CNC) word scores yet similar excellent results were seen.

CONCLUSIONS: Our study results suggest the CI should be considered when BKB scores have dropped to 85%. We suggest that rather than LVAS cases representing a challenge to cochlear implantation, they are amongst the best candidates for surgery, and should receive a CI at an earlier stage in hearing loss, when they have better speech perception. This allows stable hearing to be established earlier along with excellent speech perception outcomes.

Cochlear implants in eight children with Down Syndrome - Auditory performance and challenges in assessment.

Heldahl MG, Eksveen B, Bunne M.

OBJECTIVES: A small proportion of children with Down Syndrome (DS) have severe to profound hearing loss and may potentially benefit from a cochlear implant (CI). Evidence on outcomes in DS is very limited, and there is a need for further investigation to provide a basis for clinical evaluation of candidates and outcomes. This study aims to explore outcomes of CI in children with DS in Norway.

METHODS: Data on all children with DS and CI in Norway were collected from the CI registry and patients' records at the national pediatric CI center. Main outcome measures were: use of CI, Category of Auditory Performance (CAP), Speech Intelligibility Rate (SIR), and parents' and caregivers' views of the benefits of CI.

RESULTS: Eight children with DS have received CI in Norway, all bilaterally. The outcomes varied greatly. All children used their CIs, and all of them responded to environmental sounds. Four children reached CAP score 5 after several years of use, (i.e., they understand phrases without lip reading). All children scored at least 2, (i.e., responds to speech sounds). One child reached a SIR score of 3, (i.e. connected speech is intelligible to experienced listeners). The rest of the children reached SIR scores of 1 or 1-2, (i.e., connected speech is unintelligible). Without exception, parents had an entirely positive attitude to their children using a CI. Co-morbidity and middle ear disease frequently delayed implantation.

CONCLUSIONS: Our experience with CI in children with DS is positive. However, CI cannot replace other types of communication for these children, and it is important to give parents realistic expectations prior to surgery. Outcomes might be considered limited when evaluated with instruments for testing auditory performance and speech intelligibility constructed for children without additional disabilities. We do not believe that such outcomes reflect the benefit in real life.

Craniofacial Interventions in Children.

Hopkins B, Dean K, Appachi S, Drake AF.

Craniofacial interventions are common and the surgical options continue to grow. The issues encountered include micrognathia, macroglossia, midface hypoplasia, hearing loss, facial nerve palsy, hemifacial microsomia, and microtia. In addition, a unifying theme is complex upper airway obstruction. Throughout a child's life the focus of interventions may change from airway management to speech, hearing, and language optimization, and finally to decannulation and procedures aimed at social integration and self-esteem. Otolaryngologists play an important role in this arena and provide high-quality care while continuing to expand what can be done for our patients.

Does an early discharge of a newborn influence the success of the newborn hearing screening in developing countries? A hospital based study.

Hrnčić N, Hatibović H, Goga A, Hodžić Đ.

AIM: To investigate outcomes of newborn hearing screening (NHS) with transient evoked otoacoustic emissions (TEOAE) depending on the time from the birth to hearing screening.

METHODS: A prospective study was performed in the Cantonal Hospital Zenica, Bosnia and Herzegovina. The NHS with TEOAE was done before hospital discharge for all infants. The total of 1217 newborns were tested during a six-month period, from 1st February to 31st July 2016. The data of 1167 were available for analysis. Those data were divided in four groups depending on the time passed from the birth to hearing screening: Group A (n= 133 newborns, NHS performed in the first 24 hours after birth); group B (n = 294 newborns, NHS performed between 24-36h after birth); group C (n = 184 newborns, NHS performed between 36-48h after birth) and group D (n= 556 newborns, NHS performed later than 48h after birth).

RESULTS: Total referral rate was 19.1% (n = 223): for group A 30.1% (n=40), for group B 25.2% (n=74), for group C 19.0% (n=35) and for group D 13.3% (n=74). There was statistically significant difference between groups A and C (p=0.03), between groups A and D (p<0.001) and between groups B and D (p<0.001) in total and in well baby nursery (WBN).

CONCLUSION: The total referral rates in NHS were high because of early post birth discharge of newborns. The NHS should be performed in infants older than 36 hours according to the results in this study.

[**Restricted Speech Recognition in Noise and Quality of Life of Hearing-Impaired Children and Adolescents With Cochlear Implants - Need for Studies Addressing This Topic With Valid Pediatric Quality of Life Instruments.**](#)

[Huber M, Havas C.](#)

Cochlear implants (CI) support the development of oral language in hearing-impaired children. However, even with CI, speech recognition in noise (SRiN) is limited. This raised the question, whether these restrictions are related to the quality of life (QoL) of children and adolescents with CI and how SRiN and QoL are related to each other. As a result of a systematic literature research only three studies were found, indicating positive moderating effects between SRiN and QoL of young CI users. Thirty studies addressed the quality of life of children and adolescents with CI. Following the criteria of the World Health Organization (WHO) for pediatric health related quality of life HRQoL (1994) only a minority used validated child centered and age appropriate QoL instruments. Moreover, despite the consensus that usually children and adolescents are the most prominent informants of their own QoL (parent-reports complement the information of the children) only a minority of investigators used self-reports. Restricted SRiN may be a burden for the QoL of children and adolescents with CI. Up to now the CI community does not seem to have focused on a possible impairment of QoL in young CI users. Further studies addressing this topic are urgently needed, which is also relevant for parents, clinicians, therapists, teachers, and policy makers. Additionally investigators should use valid pediatric QoL instruments. Most of the young CI users are able to inform about their quality of life themselves

Indian J Otolaryngol Head Neck Surg. 2019 Nov;71(Suppl 2):1187-1189. doi: 10.1007/s12070-018-1256-0. Epub 2018 Jan 31.

[**Role of Otoacoustic Emissions in Hearing Assessment of Neonates: A Prospective Observational Study.**](#)

[Jadia S, Qureshi S, Raghuwanshi P, Sharma S.](#)

Hearing plays an important role for children in learning speech and language, socialization and cognitive development. The child learns to speak based on what is heard (Rundjan et al. in Sari Pediatr 6(4):149-154, 2005). The aim of the study was to find out the percentage of hearing loss by OAE among low risk and high risk neonates. All the neonates (Low and High risk) born in a tertiary care center were screened by OAE before their discharge from the hospital and after stabilizing high risk neonates. The referred neonates were followed after two weeks. Total 722 neonates were screened of which 130 were high risk and 592 were low risk. Neonates with Serum bilirubin > 20 mg/dl or requiring exchange transfusion were excluded as OAE will be unreliable in them and they should be subjected to BERA directly. Percentage of bilateral hearing loss came to be 4.2%, 4 participants were lost to follow up and percentage of hearing loss on subsequent OAE came to be 11.6%. Low birth weight was an important risk factor for hearing loss (*p* value significant on initial and follow-up). Percentage of hearing loss with low birth weight, hyperbilirubinemia, low apgar score and prematurity to be 16%. Mechanical ventilation contributed 13% of total hearing loss followed by ototoxicity. Neonates with family history of childhood SNHL, in utero infection, craniofacial anomaly and bacterial meningitis contributed 3% of total hearing loss. On subsequent follow-up, one neonate was low-risk and the other 25 were high risk, of which 3 high risks showed sustained OAE refer. However, the low risk neonate had normal outer hair cell function i.e. OAE pass. This study clearly demonstrates importance of Universal Neonatal Hearing Screening Programme so that hearing loss can be detected as early as possible and possible intervention can be taken at the earliest.

Child Care Health Dev. 2019 Sep 10. doi: 10.1111/cch.12717. [Epub ahead of print]

[**Hearing screening outcome in neonatal intensive care unit graduates from a tertiary care centre in Singapore.**](#)

[Jayagobi PA, Yeoh A, Hee KYM, Lim SB, Choo KP, Kun Kiaang HT, Lazaroo D, Daniel LM.](#)

BACKGROUND: We aimed to analyse the outcome of universal newborn hearing screening (UNHS) and high-risk hearing screening in neonatal intensive care unit (NICU) graduates in a tertiary care unit.

METHODS: The hearing screen programme comprises a 2-stage automated auditory brainstem response protocol followed by a high-risk hearing screen at 3-6 months. This study is a retrospective study of NICU graduates born between April 2002 and December 2009. Data on hearing screening, audiological assessment, and management were extracted from a computerized data management system (HITRACK).

RESULTS: Of 100,225 newborn infants, 2.9% were admitted to the NICU during the study period. The overall incidence of hearing loss (HL) of any type/severity was 35/1,000 infants. Of infants with HL, 92.4% had their first automated auditory brainstem response at/before 1 month of corrected age. The incidence of congenital permanent HL identified by the UNHS was 15.4/1,000. The corrected median age of diagnosis was 4.5 months

(1-23.5 months). Of 2,552 NICU graduates who passed the UNHS, 75.5% were retested at 3-6 months of life. Twelve infants with permanent late-onset HL were identified, raising the overall incidence of permanent HL to 19.9/1,000; 1.1/1,000 had auditory neuropathy. Of the 92 infants with HL, 89 (96.7%) had multiple risk factors.

CONCLUSIONS: There is a high incidence of HL in NICU graduates; 22.6% were late in onset. An early rescreen in those who pass the UNHS is a beneficial step for this high risk population.

Int J Pediatr Otorhinolaryngol. 2019 Sep 19;128:109691. doi: 10.1016/j.ijporl.2019.109691. [Epub ahead of print]

Hearing screening failure rate in newborn infants with hypoxic ischemic encephalopathy.

Kadioğlu Şimşek G, Kutman HGK, Canpolat FE, Büyüktiryaki M, Üstün YE.

OBJECTIVE: The objective of this study was to establish the local incidence of hearing screening failure rate in newborns with all three stages of hypoxic ischemic encephalopathy (HIE).

METHODS: This retrospective cohort study was undertaken in a tertiary neonatal intensive care unit. Medical records and hearing screening test results were collected for two years.

RESULTS: One hundred and ninety seven infants diagnosed with HIE, 20 of them died, 177 screened. Thirty five of 177 (19%) infants failed in screening test for hearing. Screening failure rate was 10/51 (19%), 20/105 (19%) and 5/21 (23%) in stage 1, 2 and 3, respectively and did not differ between HIE stages ($p=0.88$). Furthermore failure rates were similar between infants who received therapeutic hypothermia or not (20% vs 19%, $p=0.84$).

CONCLUSION: Hearing screening failure rate in HIE is quite high even in Stage 1 infants. Management and treatment of these infants should be made carefully concerning additional risks for hearing loss and long term follow-up even in Stage 1 HIE infants should be planned strictly.

J Matern Fetal Neonatal Med. 2019 Sep 8;1-10. doi: 10.1080/14767058.2019.1658732. [Epub ahead of print]

Risk factors for hearing impairment in neonates in South Africa: scoping the context for newborn hearing screening planning.

Kanji A, Khoza-Shangase K.

PURPOSE: The current study aimed to describe the medical case history factors in a group of neonates admitted to high care and Kangaroo Mother Care wards at two hospitals in Gauteng, South Africa and to explore the relationship between specific case history factors and audiological outcomes.

METHODS: This study was part of a bigger study titled “Early detection of hearing loss: exploring risk-based hearing screening within a developing country context” where all case history data were recorded from participant medical files at the time of an initial hearing screening, through the use of a case history form that was developed for this study. Results were analyzed using descriptive statistics. The relationship between case history factors and audiological outcomes was analyzed using Fisher’s exact test.

RESULTS: Findings revealed that, of all the case history data, preterm birth (95.7%), exposure to ototoxic medication (87.7%), neonatal jaundice (NNJ) (80.6%), and birthweight below 1500 g (66.7%) were the most frequently occurring case history factors in this South African sample. No statistically significant association was found between these frequently occurring case history factors and the repeat hearing screening outcomes in this sample.

CONCLUSIONS: Risk factors for hearing impairment cannot be viewed in isolation but should be considered in relation to their interaction with multiple other risk factors. Contextually understanding the profile of high risk has implications for medical intervention, as well as for the field of Audiology where planning for early identification and intervention services for hearing impairment is vital. Such contextually relevant evidence allows for a snap shot of how this population presents, what their needs and requirements are, and their possible future requirements; hence, raising important planning implications for the department of health and for targeted newborn hearing screening as well as early childhood intervention within the South African context.

Semin Hear. 2019 Aug;40(3):245-252. doi: 10.1055/s-0039-1693491. Epub 2019 Aug 13.

Gaining Greater Adherence from Patients for Amplification.

Kasewurm GA.

Every audiology curriculum features coursework and clinical practicum experiences on the knowledge and skills necessary to complete a basic audiological evaluation, diagnose the source of any abnormalities, and a plan for intervention. More advanced coursework typically includes information on balance disorders, newborn infant hearing screening, hearing conservation, amplification, and other electrophysiological tests. In addition, most programs include at least one course on the basic management principles on how to start and maintain a private practice in audiology. However, educational programs rarely include any type of coursework on how to

handle common objections to obtaining help in the form of amplification. This is rather surprising considering that more than 90% of patients whom an audiologist evaluates have sensory neural hearing loss where the primary treatment is amplification, except for people with severe hearing losses who are candidates for cochlear implants. This lack of training and experience often leaves clinicians intimidated by common objections to treatment involving purchasing amplification. This article will discuss common objections that patients voice each day to obtaining amplification and will offer possible solutions.

Orv Hetil. 2019 May;160(21):822-828. doi: 10.1556/650.2019.31398.

[Etiological factors of sensorineural hearing loss in children after cochlear implantation].

Kecskeméti N, Gáborján A, Szőnyi M, Küstel M, Baranyi I, Molnár MJ, Tamás L, Gál A, Szirmai Á.

INTRODUCTION: Congenital sensorineural hearing loss is one of the most common sensory defects affecting 1-3 children per 1000 newborns. There are a lot of causes which result in congenital hearing loss, the most common is the genetic origin, but infection, cochlear malformation or other acquired causes can be reasons as well.

AIM: The aim of this study was to establish the etiological factors of congenital profound sensorineural hearing loss in children who underwent cochlear implantation.

RESULTS: Our results show that the origin of the hearing loss was discovered in 62.9% of our patients. The most common etiological factor was the c.35delG mutation of the *gap junction protein β -2* gene, the allele frequency was 38.7% in our cohort. Infection constituted to 10.1%, and meningitis and cytomegalovirus infection were the second most common cause. 79.9% of our patients received sufficient hearing rehabilitation before the end of the speech development's period (6 years old), but 11.2% of our cases were still diagnosed late.

CONCLUSIONS: Based on our data we can state that genetic evaluation is crucial in the diagnostic process of congenital profound sensorineural hearing loss. Sufficient hearing rehabilitation affects the whole life of the child, and by late cochlear implantation the speech development falls behind. We can decrease the ratio of the late implantation with the new protocol of newborn hearing screening, and with sufficient information provided to the colleagues, so the children may be referred to the proper center for rehabilitation without delay.

J Matern Fetal Neonatal Med. 2019 Aug 11:1-5. doi: 10.1080/14767058.2019.1651277. [Epub ahead of print]

Risk of congenital cytomegalovirus infection in children born to women with IgG avidity in the grey zone during first trimester of pregnancy.

Kekkou K, Kavatha D, Karalexi M, Galani L, Dimopoulou D, Papaevangelou V, Antoniadou A.

INTRODUCTION: Cytomegalovirus (CMV) is the most common congenital viral infection and is regarded as the leading nongenetic cause of sensorineural hearing loss. Currently, international consensuses discourage prenatal screening of pregnant women. However, in few countries mainly in Southern Europe, screening of pregnant women for CMV infection is common practice. Management of women found with IgG+/IgM+ and IgG avidity titers in the grey zone during first trimester causes significant stress to both families and health care workers.

PATIENTS AND METHODS: Pregnant women referred to our outpatient clinic with the diagnosis of acute CMV infection (IgM+/IgG+) during early pregnancy (gestational age ≤ 14 weeks) and IgG avidity in the grey zone were prospectively followed. The administration of CMV-HIG was offered and follow-up included fetal U/S, amniocentesis for CMV-DNA detection and MRI when appropriate. All neonates were examined by urine PCR and prospectively followed according to existing recommendations.

RESULTS: Ninety women (mean age 30.8 years) were retrospectively analyzed. Most (79.6%) received CMV-HIG. Four women terminated pregnancy (2 unrelated to CMV reasons and 2 because of CMV-positive amniotic fluid). Eighty-seven babies were born asymptomatic. Two newborns were diagnosed with congenital CMV infection. The overall transmission rate was 4.4%; 4.3 *versus* 5.6% for those receiving or not CMV-HIG. No adverse outcomes were detected during follow-up (median 24 months). Maternal age, parity, detection of maternal CMV-viremia upon diagnosis, delay between diagnosis and consultation, gestational week of first consultation, administration of CMV-HIG and number of doses were not associated with the risk of vertical CMV transmission.

DISCUSSIONS: Vertical transmission of CMV infection in pregnancies with acute CMV-infection and IgG avidity titers in the grey zone during first trimester was 4.4%, higher than that in infants born post nonprimary infection (NPI) during pregnancy. More powered studies are needed to prove a significant reduction in transmission using CMV-HIG.

J Laryngol Otol. 2019 May;133(5):363-367. doi: 10.1017/S0022215119000872. Epub 2019 May 9.

[Accuracy of otoacoustic emissions, and automated and diagnostic auditory brainstem responses, in high-risk infants.](#)

[Khaimook W, Pantuyosyanyong D, Pitathawatchai P.](#)

OBJECTIVES: This study aimed to compare the diagnostic reliabilities of transient evoked otoacoustic emissions, automated auditory brainstem responses and brainstem auditory evoked responses for detecting hearing loss, and to use the information regarding hearing level of automated auditory brainstem responses for planning rehabilitation.

METHODS: A total of 144 high-risk infants (288 ears) completed the 3 hearing tests. The sensitivity and specificity of otoacoustic emissions and automated auditory brainstem responses were compared using the chi-square test.

RESULTS: Automated auditory brainstem response was the most reliable test of hearing levels, with a sensitivity of 91.7 per cent and specificity of 92.1 per cent; the sensitivity of otoacoustic emissions was 78.7 per cent and the specificity was 88.8 per cent.

CONCLUSION: Automated auditory brainstem responses have acceptably high sensitivity and specificity. Additionally, the hearing level from automated auditory brainstem responses can help the screeners explain to the parents the importance of further diagnosis and rehabilitation.

Ann Otol Rhinol Laryngol. 2019 Jun;128(6_suppl):61S-68S. doi: 10.1177/0003489419835234.

[Clinical Value of Measurement of Internal Auditory Canal in Pediatric Cochlear Implantation.](#)

[Kim H, Kim DY, Ha EJ, Park HY.](#)

OBJECTIVES: The aims of this study were to clarify the clinical value of the bony cochlear nerve canal (BCNC) and internal auditory canal (IAC) in children with bilateral sensorineural hearing loss (b-SNHL) and to reveal the correlation between these parameters and outcomes after cochlear implantation (CI).

METHODS: Ninety-four ears with b-SNHL that received CI and 100 ears with normal hearing were enrolled. Parameters of IAC and pre- and post-CI categories of auditory performance scores were analyzed.

RESULTS: The width of the BCNC and the width, height, and length of the IAC were shorter in the b-SNHL group. BCNC and IAC width were associated with b-SNHL. The calculated cutoff values for BCNC and IAC width were 2.055 mm in the BCNC and 4.245 mm in the IAC, setting the sensitivity to 90%. Patients with narrow BCNCs and IACs had significantly worse post-CI auditory performance.

CONCLUSIONS: BCNC and IAC widths were narrower in children with b-SNHL than in normal-hearing children. Narrow BCNC and IAC width had a negative impact on post-CI outcomes. The proposed cutoff values for BCNC and IAC width were meaningful when predicting the auditory outcome after CI, especially considering both.

Front Cell Infect Microbiol. 2019 Oct 4;9:339. doi: 10.3389/fcimb.2019.00339. eCollection 2019.

[Altered Middle Ear Microbiome in Children With Chronic Otitis Media With Effusion and Respiratory Illnesses.](#)

[Kolbe AR, Castro-Nallar E, Preciado D, Pérez-Losada M.](#)

Chronic otitis media with effusion (COME) is a common childhood disease characterized by an accumulation of fluid behind the eardrum. COME often requires surgical intervention and can also lead to significant hearing loss and subsequent learning disabilities. Recent characterization of the middle ear fluid (MEF) microbiome in pediatric patients has led to an improved understanding of the microbiota present in the middle ear during COME. However, it is not currently known how the MEF microbiome might vary due to other conditions, particularly respiratory disorders. Here, we apply an amplicon sequence variant (ASV) pipeline to MEF 16S rRNA high-throughput sequencing data from 50 children with COME (ages 3-176 months) undergoing tube placement. We achieve a more detailed taxonomic resolution than previously reported, including species and genus level resolution. Additionally, we provide the first report of the functional roles of the MEF microbiome and demonstrate that despite high taxonomic diversity, the functional capacity of the MEF microbiome remains uniform between patients. Furthermore, we analyze microbiome differences between children with COME with and without a history of lower airway disease (i.e., asthma or bronchiolitis). The MEF microbiome was less diverse in participants with lower airway disease than in patients without, and phylogenetic β -diversity (weighted UniFrac) was significantly different based on lower airway disease status. Differential abundance between patients with lower airway disease and those without was observed for the genera *Haemophilus*, *Moraxella*, *Staphylococcus*, *Alloiococcus*, and *Turicella*. These findings support previous suggestions of a link between COME and respiratory illnesses and emphasize the need for future study of the middle ear and respiratory tract microbiomes in diseases such as asthma and bronchiolitis.

Newborn Hearing Screening: Early Education = More Satisfied Mothers.

Krishnan LA, Van Hyfte S, Richards KAR.

Newborn hearing screening (NBHS) has become the norm in all states in the United States. However, parents receive limited information about it, usually at the hospital, and have low awareness about the process. Yet parents and professionals agree that communication about the NBHS process should begin before childbirth (Arnold et al., 2006). Having information about the screening process, simply by being present for the screening and knowing the results, has been shown to positively affect parental attitudes toward NBHS (Weichbold, Welzl-Mueller, & Mussbacher, 2001).

Purpose: The aim of this study was to assess whether providing expectant mothers with information related to the NBHS process in the 3rd trimester will increase their satisfaction with the NBHS program. **Method:** Partnering with a local obstetrician, expectant mothers were provided with information regarding NBHS. A modified version of the Parent Satisfaction Questionnaire with Neonatal Hearing Screening Program (Mazlan, Hickson, & Driscoll, 2006) was administered to mothers who received information and mothers who did not receive any education.

Results: Quantitative results indicate significantly increased satisfaction with the NBHS program when early education was received. Qualitative comments from participants support the quantitative findings. **Conclusion:** Results suggest that educating obstetricians and their nurses about NBHS and having them provide information to expectant mothers, as well as having mothers present for the screening, may increase parental satisfaction and therefore benefit early hearing detection and intervention programs.

J Laryngol Otol. 2019 Aug 22:1-9. doi: 10.1017/S0022215119001658. [Epub ahead of print]

Paediatric hearing loss: a community-based survey in peri-urban Kumasi, Ghana.

Larsen-Reindorf R, Otopiri E, Anomah JE, Edwards BM, Frimpong B, Waller B, Prince ME, Basura GJ.

BACKGROUND: Paediatric hearing loss rates in Ghana are currently unknown.

METHODS: A cross-sectional study was conducted in peri-urban Kumasi, Ghana; children (aged 3-15 years) were recruited from randomly selected households. Selected children underwent otoscopic examination prior to in-community pure tone screening using the portable ShoeBox audiometer. The LittleEars auditory questionnaire was also administered to caregivers and parents.

RESULTS: Data were collected from 387 children. After conditioning, 362 children were screened using monaural pure tones presented at 25 dB. Twenty-five children could not be conditioned to behavioural audiometric screening. Eight children were referred based on audiometric screening results. Of those, four were identified as having hearing loss. Four children scored less than the maximum mark of 35 on the LittleEars questionnaire. Of those, three had hearing loss as identified through pure tone screening. The predominant physical finding on otoscopy was ear canal cerumen impaction.

CONCLUSION: Paediatric hearing loss is prevalent in Ghana, and should be treated as a public health problem warranting further evaluation and epidemiology characterisation.

Otol Neurotol. 2019 Aug;40(7):927-935. doi: 10.1097/MAO.0000000000002299.

Endaural Laser-Assisted Single-Stage Inside-Out Cholesteatoma Surgery (LASIC) to Treat Advanced Congenital Cholesteatoma.

Lee CH, Kim MK, Kim HM, Won C, Shin TH, Kim SY.

OBJECTIVES: Investigation of endaural laser-assisted single-stage inside-out cholesteatoma surgery (LASIC) to treat advanced congenital cholesteatoma (ACC) by a modified staging system based on ossicle status.

STUDY DESIGN: A retrospective case review.

SETTING: A university hospital otology referral clinic.

PATIENTS: Two hundred consecutive pediatric patients with ACC were enrolled.

INTERVENTIONS: Endaural LASIC and postoperative temporal bone computed tomography (CT).

MAIN OUTCOME MEASURES: Residual or recurrent CC and audiological outcomes.

RESULTS: LASIC was feasible in 98.0% of patients. Single-stage ossiculoplasty was performed in 95.5% of patients. Hearing preservation to less than 20dB was 59.2% and to the preoperative hearing level was 84.5%. Mastoid invasion did not result in worse recidivism or hearing loss (HL) but further invasion of the stapes superstructure (stage IV) significantly elevated both the recidivism (16.7%) and the risk for HL (to 84.8%) ($p=0.001$). Ossicle preservation LASIC was frequently possible in stage III posterior type (75.6%), whereas it was rarely possible (15.4%) in the anterior type. Incudostapedial joint (ISJ) invasion in the absence of cochleariform process (CP) invasion (III-posterior) did not increase the incidence of HL (6.1%) or recidivism (2.4%). However,

simultaneous invasion of the CP and ISJ (III-anterior) elevated the risk of HL by 46.2% by ossicle removal, although recidivism was not increased (3.8%).

CONCLUSIONS: Endaural LASIC for ACC achieved satisfactory recidivism (overall 7.5%, 16.7% in stage IV) comparable to early CC (17.2%). An ossicle status-based staging system was more efficient for correlation with audiologic and surgical outcomes of CC than that of mastoid invasion.

Otol Neurotol. 2019 Oct;40(9):e909-e917. doi: 10.1097/MAO.0000000000002339.

Lessons From an Analysis of Newborn Hearing Screening Data for Children With Cochlear Implants.

Lee JM, Lee HJ, Jung J, Moon IS, Kim SH, Kim J, Choi JY.

OBJECTIVES: The aims of the study are to identify the limitations of the current newborn hearing screening (NHS) programs and provide recommendations for better protocols.

STUDY DESIGN: Retrospective analysis.

SETTING: Tertiary referral center.

PATIENTS: The study participants were 185 children who received cochlear implants (CIs) at ≤ 5 years of age.

INTERVENTIONS: Therapeutic and rehabilitative.

MAIN OUTCOME MEASURES: The results of NHS, screening tools used, age, and hearing thresholds at which hearing loss was confirmed, causes of the hearing loss, age of CI insertion, aided pure-tone audiogram findings, and language development level were analyzed.

RESULTS: NHS data was available for 109 children, and 24 patients (22.0%) had passed NHS for both ears. Hearing loss was confirmed considerably later in children who had passed NHS than in children who were referred for further evaluation ($p < 0.01$). The most common cause for hearing loss in the NHS-pass group was SLC26A4 mutations (41.7%). Patients in the NHS-pass group received CIs considerably later than those in the NHS-referred group ($p < 0.01$). Among patients with SLC26A4 mutations, the language development level was significantly lower in the NHS-pass group than in the NHS-referred group ($p < 0.01$).

CONCLUSIONS: Careful counseling regarding NHS results is necessary for parents to understand that the absence of hearing loss at birth does not mean that the child will not develop hearing loss later in life. Genetic testing for SLC26A4 mutations may be necessary in regions with a high incidence of these mutations, such as East Asia.

Int J Pediatr Otorhinolaryngol. 2019 Oct 11;128:109720. doi: 10.1016/j.ijporl.2019.109720. [Epub ahead of print]

Trajectory of auditory and language development in the early stages of pre-lingual children post cochlear implantation: A longitudinal follow up study.

Li G, Zhao F, Tao Y, Zhang L, Yao X, Zheng Y.

OBJECTIVES: The aim of this longitudinal follow-up study was to explore the trajectories of early auditory and language development in Mandarin speaking children younger than 3 years of age following switch-on of their cochlear implants (CIs).

METHODS: Early auditory and language development was measured longitudinally using the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS), which is a commonly used tool for assessing early prelingual auditory development (EPLAD) in children, and the subtest (Words and Gestures, W&G) of the simplified short form version of the Mandarin Communicative Development Inventory (SSF-MCDI) to assess receptive and expressive vocabulary growths of children in 24 pediatric cochlea implant recipients at baseline, 3, 6, and 12 months following switch-on. Age at switch-on ranged from 1 to 3 years of age. Participants were divided into two groups based on age at switch-on. The IT-MAIS and SSF-MCDI (W&G) scores were analyzed with comparison to normal children, unaided hearing-impaired children, and CI children.

RESULTS: Significant improvements in IT-MAIS and SSF-MCDI (W&G) scores from baseline to 12 months were seen after switch-on in both CI groups and were comparable to the normal hearing children in the first year of age. The IT-MAIS scores of CI children in both groups at 12 months after switch-on surpassed the average level of unaided peers with profound hearing loss and were similar to the average level of unaided peers with mild hearing loss. SSF-MCDI (W&G) scores in word comprehension and expression were significantly different between groups at some intervals.

CONCLUSIONS: Children younger than 3 years of age with cochlear implants have similar trajectories in early auditory and language developments to normally hearing children. Moreover, early implantation is an important factor for the early auditory development when comparing EPLAD results between CI children and unaided peers with different hearing loss. Finally, it is noteworthy that CI children master the skill of word comprehension before the skill of word expression, and that word comprehension may be the basis of word expression.

[Contribution of the GSTP1 c.313A>G variant to hearing loss risk in patients exposed to platinum chemotherapy during childhood.](#)

[Lieberman PHP](#), [Goffi-Gomez MVS](#), [Schultz C](#), [Jacob PL](#), [de Paula CAA](#), [Sartorato EL](#), [Torrezan GT](#), [Ferreira EN](#), [Carraro DM](#).

BACKGROUND AND AIM: Ototoxicity is a potential adverse effect of chemotherapy with platinum drugs, such as cisplatin and carboplatin, in children. Hearing loss (HL) affecting frequencies below 4 kHz can compromise speech perception. The aim of this study was to investigate whether genetic variants previously implicated in ototoxicity are associated with HL overall and HL below 4 kHz in pediatric oncology patients treated with cisplatin or carboplatin.

MATERIALS AND METHODS: Patients given cisplatin or carboplatin for a pediatric cancer at least 5 years prior to the start of the study were enrolled. The patients underwent comprehensive audiological evaluations and genotyping to detect the presence of the GJB2 c.35delG, GSTP1 c.313A>G, and MT-RNR1 m.1555A>G polymorphisms.

RESULTS: HL was identified in 31/61 patients (50.8%), including 28/42 treated with cisplatin (66.6%) and 3/19 treated with carboplatin (15.8%). HL was associated with higher mean doses of cisplatin ($p = .002$) and carboplatin ($p = .010$). The c.313A>G variant of GSTP1 (heterozygous or homozygous) was detected in 31/61 patients (50.8%). An association between this variant allele and HL involving frequencies ≤ 4 kHz was identified ($p = .020$; 10-fold vs. non-carriers). No associations with HL were observed for GJB2 or MT-RNR1 gene variants.

CONCLUSION: The GSTP1 c.313A>G variant may increase the risk of low-frequency HL in pediatric oncology patients treated with cisplatin or carboplatin chemotherapy.

Laryngoscope. 2019 May 27. doi: 10.1002/lary.28095. [Epub ahead of print]

[Imaging findings in pediatric single-sided deafness and asymmetric hearing loss.](#)

[Lipschitz N](#), [Kohlberg GD](#), [Scott M](#), [Greinwald JH Jr](#).

OBJECTIVE: To examine the imaging findings on computer tomography (CT) and magnetic resonance imaging (MRI) in pediatric single-sided deafness (SSD) and asymmetric hearing loss (ASH).

METHODS: The medical records of 189 pediatric patients with SSD and ASH were retrospectively reviewed, and imaging findings were compared. SSD was defined as unilateral profound hearing loss and contralateral normal hearing ear. In the ASH group, ASHw was defined as the worse hearing ear with profound hearing loss, while ASHb was defined as the better hearing ear with mild-moderate hearing loss.

RESULTS: There were 170 patients with SSD and 19 patients with ASH. In the SSD group, 83 patients (48.8%) had imaging findings associated with hearing loss. In the ASH group, such imaging findings were found in six (31.6%) of the ASHw and in five (26.3%) of the ASHb ears. The most common finding in the SSD group was cochlear nerve deficiency (50.6%), followed by cochlear dysplasia (39.8%) and enlarged vestibular aqueduct (26.5%). In the ASH groups, cochlear dysplasia was seen in three (50%) of ASHw ears and in two (40%) of the ASHb ears, and enlarged vestibular aqueduct was seen in three (50%) of ASHw ears and in two (40%) of the ASHb ears.

CONCLUSION: Imaging studies identified the etiology in half of the cases of SSD and in one-third of ASH patients. Our findings strongly support the use of imaging studies in the evaluation of pediatric SSD and ASH.

LEVEL OF EVIDENCE: 4 Laryngoscope, 2019.

Ann Agric Environ Med. 2019 Sep 19;26(3):445-449. doi: 10.26444/aaem/102691. Epub 2019 Mar 15.

[Effectiveness of health-promoting activities in the area of general hearing screening tests in newborns in Poland.](#)

[Lisiecka-Bielanowicz M](#), [Molenda BA](#).

INTRODUCTION: An analysis of the impact of health promotion efforts as part of the Polish Universal Neonatal Hearing Screening Program demonstrated diagnostic efficacy. The health promotion efforts had been efficiently implemented via a proper execution of this long-term Program by the Great Orchestra for Christmas Charity (WOŚP) foundation.

OBJECTIVE: The aim of the study was to demonstrate the impact of health promotion efforts by public benefit organizations by analyzing the WOŚP foundation's involvement in the Program in Poland during 2003-2015.

MATERIAL AND METHODS: Statistical data for 2003-2015 were obtained from the database of the Department of Otolaryngology and Laryngeal Oncology at Poznan University of Medical Sciences. The analyzed data included the rates of hearing-impairment detection (phase 1), diagnosis (phase 2), and treatment (phase 3) in the newborns included in the Program, as well as the documentation of expenditures based on WOŚP's foundation annual financial reports.

RESULTS:

A total of 4,672,704 newborns were screened from 2003-2015, which amounted to approximately 360,000 screened patients per year. During the analyzed 12 years of the Program, the mean proportion of children who underwent screening was 96.1%, with an observable, gradual increase in the number of children undergoing diagnostic assessments (phase 2) for a suspected hearing impairment, with the largest amount of money having been allocated to the Program implementation and expended in its early stages. The total cost of financing the Program by the year 2015 was 51,841,712 PLN.

CONCLUSIONS: The Polish Universal Neonatal Hearing Screening Program resulted in an improvement in the quality in the screening test, ensured more thorough diagnostics, and shortened the time needed to implement the appropriate treatment.

Otolaryngol Clin North Am. 2019 Oct;52(5):847-861. doi: 10.1016/j.otc.2019.05.004. Epub 2019 Jul 12.

[Advances in Management of Pediatric Sensorineural Hearing Loss.](#)

[Liu CC](#), [Anne S](#), [Horn DL](#).

The work-up and management of sensorineural hearing loss in children has been an area of rapid evolution. With the availability of genetic and cytomegalovirus testing, the diagnostic process is continuously refined. Aural rehabilitation should be provided to children in a timely manner. At present, the main surgical options for the treatment of sensorineural hearing loss are bone conduction sound processors and cochlear implants. Investigations into modalities such as auditory brainstem implants are ongoing. With further technological and medical advancements, the evaluation and management of pediatric sensorineural hearing loss will undoubtedly continue to change.

Ear Hear. 2019 May 20. doi: 10.1097/AUD.0000000000000744. [Epub ahead of print]

[The Effects of GJB2 or SLC26A4 Gene Mutations on Neural Response of the Electrically Stimulated Auditory Nerve in Children.](#)

[Luo J](#), [Xu L](#), [Chao X](#), [Wang R](#), [Pellitteri A](#), [Bai X](#), [Fan Z](#), [Wang H](#), [He S](#).

OBJECTIVES: This study aimed to (1) investigate the effect of GJB2 and SLC26A4 gene mutations on auditory nerve function in pediatric cochlear implant users and (2) compare their results with those measured in implanted children with idiopathic hearing loss.

DESIGN: Participants included 20 children with biallelic GJB2 mutations, 16 children with biallelic SLC26A4 mutations, and 19 children with idiopathic hearing loss. All subjects except for two in the SLC26A4 group had concurrent Mondini malformation and enlarged vestibular aqueduct. All subjects used Cochlear Nucleus devices in their test ears. For each subject, electrophysiological measures of the electrically evoked compound action potential (eCAP) were recorded using both anodic- and cathodic-leading biphasic pulses. Dependent variables (DVs) of interest included slope of eCAP input/output (I/O) function, the eCAP threshold, and eCAP amplitude measured at the maximum comfortable level (C level) of the anodic-leading stimulus (i.e., the anodic C level). Slopes of eCAP I/O functions were estimated using statistical modeling with a linear regression function. These DVs were measured at three electrode locations across the electrode array. Generalized linear mixed effect models were used to evaluate the effects of study group, stimulus polarity, and electrode location on each DV.

RESULTS: Steeper slopes of eCAP I/O function, lower eCAP thresholds, and larger eCAP amplitude at the anodic C level were measured for the anodic-leading stimulus compared with the cathodic-leading stimulus in all subject groups. Children with GJB2 mutations showed steeper slopes of eCAP I/O function and larger eCAP amplitudes at the anodic C level than children with SLC26A4 mutations and children with idiopathic hearing loss for both the anodic- and cathodic-leading stimuli. In addition, children with GJB2 mutations showed a smaller increase in eCAP amplitude when the stimulus changed from the cathodic-leading pulse to the anodic-leading pulse (i.e., smaller polarity effect) than children with idiopathic hearing loss. There was no statistically significant difference in slope of eCAP I/O function, eCAP amplitude at the anodic C level, or the size of polarity effect on all three DVs between children with SLC26A4 mutations and children with idiopathic hearing loss. These results suggested that better auditory nerve function was associated with GJB2 but not with SLC26A4 mutations when compared with idiopathic hearing loss. In addition, significant effects of electrode location were observed for slope of eCAP I/O function and the eCAP threshold.

CONCLUSIONS: GJB2 and SLC26A4 gene mutations did not alter polarity sensitivity of auditory nerve fibers to electrical stimulation. The anodic-leading stimulus was generally more effective in activating auditory nerve fibers than the cathodic-leading stimulus, despite the presence of GJB2 or SLC26A4 mutations. Patients with GJB2 mutations appeared to have better functional status of the auditory nerve than patients with SLC26A4 mutations who had concurrent Mondini malformation and enlarged vestibular aqueduct and patients with idiopathic hearing loss.

Long-term follow-up of auditory performance and speech perception and effects of age on cochlear implantation in children with pre-lingual deafness.

Lyu J, Kong Y, Xu TQ, Dong RJ, Qi BE, Wang S, Li YX, Liu HH, Chen XQ.

BACKGROUND: The development of auditory and speech perception ability of children with hearing loss is affected by many factors after they undergo cochlear implantation (CI). Age at CI (CI age) appears to play an important role among these factors. This study aimed to evaluate the development of auditory and speech perception ability and explore the impact of CI age on children with pre-lingual deafness present before 3 years of age.

METHODS: Two hundred and seventy-eight children with pre-lingual deafness (176 boys and 102 girls) were included in this study, and the CI age ranged from 6 to 36 months (mean age, 19 months). Categorical auditory performance (CAP) was assessed to evaluate auditory ability, and the speech intelligibility rating was used to evaluate speech intelligibility. The evaluations were performed before CI and 1, 3, 6, 12, 18, 24, 36, 48, and 60 months after CI.

RESULTS: The auditory ability of the pre-lingually hearing-impaired children showed the fastest development within 6 months after CI ($k=0.524$, $t=30.992$, $P<0.05$); then, the progress started to decelerate ($k=0.14$, $t=3.704$, $P<0.05$) and entered a plateau at the 24th month ($k=0.03$, $t=1.908$, $P<0.05$). Speech intelligibility showed the fastest improvement between the 12th and 24th months after CI ($k=0.138$, $t=5.365$, $P<0.05$); then, the progress started to decelerate ($k=0.026$, $t=1.465$, $P<0.05$) and entered a plateau at the 48th month ($k=0.012$, $t=1.542$, $P<0.05$). The CI age had no statistical significant effect on the auditory and speech abilities starting at 2 years after CI ($P>0.05$). The optimal cutoff age for CI was 15 months.

CONCLUSIONS: Within 5 years after CI, the auditory and speech ability of young hearing-impaired children continuously improved, although speech development lagged behind that of hearing. An earlier CI age is recommended; the optimal cutoff age for CI is at 15 months.

Active transcutaneous bone conduction hearing implants: Systematic review and meta-analysis.

Magele A, Schoerg P, Stanek B, Gradl B, Sprinzi GM.

BACKGROUND: In July 2018 the active transcutaneous bone conduction hearing implant received FDA approval in the US (for patients 12 years and older with conductive and/or mixed hearing loss or single-sided deafness), reflecting the current trend of moving away from percutaneous hearing solutions towards intact skin systems.

OBJECTIVES: To critically assess the current literature on safety, efficacy and subjective benefit after implantation with an active transcutaneous bone conduction hearing device.

DATA SOURCES: Literature investigation was performed by electronic database search including PubMed and Cochrane Central Register of Controlled Trials, and manual search of relevant journals and reference lists of included studies.

STUDY ELIGIBILITY CRITERIA: Randomized controlled trials, clinical controlled trials and cohort studies, case series and case reports investigating subjective and objective outcomes.

STUDY APPRAISAL AND SYNTHESIS METHODS: Retrieved literature was screened and extracted by two reviewers independently. Subgroup analysis of indications (conductive and/or mixed hearing loss, single-sided deafness) and participant ages (pediatric vs. adults) was conducted on patients with active transcutaneous bone conduction devices. Sensitivity analysis was performed to test the stability of the results in meta-analysis.

RESULTS: 39 citations reporting on pre- and postoperative audiological results, speech performance in quiet and in noise, localization testing as well as subjective outcomes were included in this systematic review. Functional gain as well as word recognition score outcomes could be further investigated via meta-analysis. All outcomes reported and summarized here reflect beneficial audiological performance and high patient satisfaction, accompanied with a low complications rate (minor event incidence rate: 9.9 person-years; major incidence rate: 148.9 person-years) for the indications of conductive and mixed hearing loss as well as in individuals suffering from single-sided deafness for all age groups of subjects who underwent active transcutaneous bone conduction hearing device implantation.

LIMITATIONS: A limiting factor of this systematic review was the Level of Evidence of the reviewed literature, comprising 2a/3a studies (cohort studies and case-control studies). Furthermore, the reporting standards, especially in outcomes such as word recognition scores in quiet and in noise, vary across study cites from various countries, which impedes comparisons. Last but not least, no other comparable other device was retrieved as the active transcutaneous bone conduction hearing device is the only available at the moment.

CONCLUSION: The device's transcutaneous technology results in a minor event incidence rate of one in 9.9 person-years and a major incidence rate of one in 148.9 person-years. Based on the audiological outcomes, high

patient satisfaction as well as the low complication rate, the authors recommend the active transcutaneous bone conduction hearing device as a safe and effective treatment for patients suffering from hearing loss within the device's indication criteria (conductive and/or mixed hearing loss or single-sided deafness).

J Child Health Care. 2019 Jun;23(2):232-241. doi: 10.1177/1367493518788477. Epub 2018 Aug 1.

[Hearing impairment detection and intervention in children from centre-based early intervention programmes.](#)

[Maluleke NP](#), [Khoza-Shangase K](#), [Kanji A](#).

The study aimed to describe ages at identification and initiation of early intervention (EI) services for children enrolled in centre-based EI programmes in Gauteng, as well as to describe the nature of EI services that the children received. The researchers conducted retrospective record reviews of the EI programme files. In addition, caregivers of eight children identified with hearing impairments and enrolled in centre-based EI programmes in Gauteng completed a newly constructed questionnaire. The caregiver questionnaire produced data pertaining to the child's family demographics, background information and schooling history. Descriptive statistics were used to analyse the data, using frequency distribution and measures of central tendency. None of the children received newborn hearing screening services, thus they were identified late following maternal suspicion of hearing impairment. Late identification of the hearing impairment resulted in suboptimal initiation of EI services. All the children received aural habilitation and/or speech-language therapy services. These findings indicate that there is a great need for the establishment of widespread early hearing detection and intervention programmes that will lead to earlier identification of infant and childhood hearing impairment and timely initiation of EI services.

J Inherit Metab Dis. 2019 Jul;42(4):581-597. doi: 10.1002/jimd.12125. Epub 2019 Jun 24.

[Genetic defects of thiamine transport and metabolism: A review of clinical phenotypes, genetics, and functional studies.](#)

[Marcé-Grau A](#), [Martí-Sánchez L](#), [Baide-Mairena H](#), [Ortigoza-Escobar JD](#), [Pérez-Dueñas B](#).

Thiamine is a crucial cofactor involved in the maintenance of carbohydrate metabolism and participates in multiple cellular metabolic processes within the cytosol, mitochondria, and peroxisomes. Currently, four genetic defects have been described causing impairment of thiamine transport and metabolism: SLC19A2 dysfunction leads to diabetes mellitus, megaloblastic anemia and sensory-neural hearing loss, whereas SLC19A3, SLC25A19, and TPK1-related disorders result in recurrent encephalopathy, basal ganglia necrosis, generalized dystonia, severe disability, and early death. In order to achieve early diagnosis and treatment, biomarkers play an important role. SLC19A3 patients present a profound decrease of free-thiamine in cerebrospinal fluid (CSF) and fibroblasts. TPK1 patients show decreased concentrations of thiamine pyrophosphate in blood and muscle. Thiamine supplementation has been shown to improve diabetes and anemia control in Rogers' syndrome patients due to SLC19A2 deficiency. In a significant number of patients with SLC19A3, thiamine improves clinical outcome and survival, and prevents further metabolic crisis. In SLC25A19 and TPK1 defects, thiamine has also led to clinical stabilization in single cases. Moreover, thiamine supplementation leads to normal concentrations of free-thiamine in the CSF of SLC19A3 patients. Herein, we present a literature review of the current knowledge of the disease including related clinical phenotypes, treatment approaches, update of pathogenic variants, as well as in vitro and in vivo functional models that provide pathogenic evidence and propose mechanisms for thiamine deficiency in humans.

Am J Speech Lang Pathol. 2019 Aug 9;28(3):1318-1334. doi: 10.1044/2019_AJSLP-18-0160. Epub 2019 Jun 28.

[Effects of Age at Cochlear Implantation on Learning and Cognition: A Critical Assessment.](#)

[Marschark M](#), [Duchesne L](#), [Pisoni D](#).

PURPOSE: Age at cochlear implantation frequently is assumed to be a key predictor of pediatric implantation benefits, but outcomes related to learning and cognition appear inconsistent. This critical assessment examines relevant literature in an effort to evaluate the impact of age at implantation in those domains for individuals who received their devices as children.

METHOD: We examined 44 peer-reviewed articles from 2003 to 2018 considering age at implantation and conducted statistical analyses regarding its impact on several domains, including literacy, academic achievement, memory, and theory of mind.

RESULTS: Across 167 assessments in various experiments and conditions, only 21% of the analyses related to age at implantation yielded evidence in favor of earlier implantation, providing greater benefits to academic achievement, learning, or cognition compared to implantation later in childhood. Among studies that considered cognitive processing (e.g., executive function, memory, visual-spatial functioning), over twice as many analyses indicated significant benefits of earlier implantation when it was considered as a discrete rather than a continuous variable.

CONCLUSION: Findings raise methodological, practical, and theoretical questions concerning how “early” is defined in studies concerning early cochlear implantation, the impact of confounding factors, and the use of nonstandard outcome measures. The present results and convergent findings from other studies are discussed in terms of the larger range of variables that need to be considered in evaluating the benefits of cochlear implantation and question the utility of considering age at implantation as a “gold standard” with regard to evaluating long-term outcomes of the procedure as a medical treatment/intervention for hearing loss.

Int J Pediatr Otorhinolaryngol. 2019 Oct;125:92-97. doi: 10.1016/j.ijporl.2019.06.028. Epub 2019 Jun 28.

[Comparison of the prevalence and features of inner ear malformations in congenital unilateral and bilateral hearing loss.](#)

[Masuda S](#), [Usui S](#).

OBJECTIVES: The aim of the study was to clarify differences in the prevalence and features of bony malformations in inner ear between congenital unilateral sensorineural hearing loss (USNHL) and congenital bilateral sensorineural hearing loss (BSNHL).

METHODS: We conducted a retrospective study of 378 consecutive infants referred from routine newborn hearing screening in the past 18 years. Clinical background, audiological data, and temporal bone computed tomography (CT) findings were analyzed. The prevalence of malformations between USNHL and BSNHL groups were compared using the Chi-square test.

RESULTS: The proportion of family history of hearing loss was significantly higher in infants with BSNHL than in those with USNHL (26/107 [24.3%] vs. 4/105 [3.7%]; $p=0.0001$). Temporal bone CT scanning revealed significantly a higher prevalence of inner ear malformations in infants with USNHL than in those with BSNHL (93/109 [85.3%] vs. 4/107 [3.7%]; $p<0.0001$). The most frequent anomaly in USNHL was cochlear nerve canal stenosis (69.7%), followed by cochlear malformations (20.2%), and narrow internal auditory canal (17.4%). Four infants with BSNHL accompanied by inner ear anomaly had complications such as Down’s syndrome, developmental delay, or epilepsy.

CONCLUSIONS: The prevalence of bony malformations in inner ear and/or IAC was markedly higher in infants with congenital USNHL than in infants with BSNHL. Temporal bone CT scanning may help to clarify the etiology of congenital hearing loss, especially in USNHL.

Front Neurosci. 2019 Oct 15;13:1093. doi: 10.3389/fnins.2019.01093. eCollection 2019.

[Auditory, Cognitive, and Linguistic Factors Predict Speech Recognition in Adverse Listening Conditions for Children With Hearing Loss.](#)

[McCreery RW](#), [Walker EA](#), [Spratford M](#), [Lewis D](#), [Brennan M](#).

OBJECTIVES: Children with hearing loss listen and learn in environments with noise and reverberation, but perform more poorly in noise and reverberation than children with normal hearing. Even with amplification, individual differences in speech recognition are observed among children with hearing loss. Few studies have examined the factors that support speech understanding in noise and reverberation for this population. This study applied the theoretical framework of the Ease of Language Understanding (ELU) model to examine the influence of auditory, cognitive, and linguistic factors on speech recognition in noise and reverberation for children with hearing loss.

DESIGN: Fifty-six children with hearing loss and 50 age-matched children with normal hearing who were 7-10 years-old participated in this study. Aided sentence recognition was measured using an adaptive procedure to determine the signal-to-noise ratio for 50% correct (SNR50) recognition in steady-state speech-shaped noise. SNR50 was also measured with noise plus a simulation of 600 ms reverberation time. Receptive vocabulary, auditory attention, and visuospatial working memory were measured. Aided speech audibility indexed by the Speech Intelligibility Index was measured through the hearing aids of children with hearing loss.

RESULTS: Children with hearing loss had poorer aided speech recognition in noise and reverberation than children with typical hearing. Children with higher receptive vocabulary and working memory skills had better speech recognition in noise and noise plus reverberation than peers with poorer skills in these domains. Children with hearing loss with higher aided audibility had better speech recognition in noise and reverberation than peers with poorer audibility. Better audibility was also associated with stronger language skills.

CONCLUSIONS: Children with hearing loss are at considerable risk for poor speech understanding in noise and in conditions with noise and reverberation. Consistent with the predictions of the ELU model, children with stronger vocabulary and working memory abilities performed better than peers with poorer skills in these domains. Better aided speech audibility was associated with better recognition in noise and noise plus reverberation conditions for children with hearing loss. Speech audibility had direct effects on speech recognition in noise and reverberation and cumulative effects on speech recognition in noise through a positive association with language development over time.

[Listening Difficulties in Children With Fetal Alcohol Spectrum Disorders: More Than a Problem of Audibility.](#)

[McLaughlin SA](#), [Thorne JC](#), [Jirikowic T](#), [Waddington T](#), [Lee AKC](#), [Astley Hemingway SJ](#).

PURPOSE: Data from standardized caregiver questionnaires indicate that children with fetal alcohol spectrum disorders (FASDs) frequently exhibit atypical auditory behaviors, including reduced responsivity to spoken stimuli. Another body of evidence suggests that prenatal alcohol exposure may result in auditory dysfunction involving loss of audibility (i.e., hearing loss) and/or impaired processing of clearly audible, “suprathreshold” sounds necessary for sound-in-noise listening. Yet, the nexus between atypical auditory behavior and underlying auditory dysfunction in children with FASDs remains largely unexplored. Method To investigate atypical auditory behaviors in FASDs and explore their potential physiological bases, we examined clinical data from 325 children diagnosed with FASDs at the University of Washington using the FASD 4-Digit Diagnostic Code. Atypical behaviors reported on the “auditory filtering” domain of the Short Sensory Profile were assessed to document their prevalence across FASD diagnoses and explore their relationship to reported hearing loss and/or central nervous system measures of cognition, attention, and language function that may indicate suprathreshold processing deficits.

RESULTS: Atypical auditory behavior was reported among 80% of children with FASDs, a prevalence that did not vary by FASD diagnostic severity or hearing status but was positively correlated with attention-deficit/hyperactivity disorder. In contrast, hearing loss was documented in the clinical records of 40% of children with fetal alcohol syndrome (FAS; a diagnosis on the fetal alcohol spectrum characterized by central nervous system dysfunction, facial dysmorphism, and growth deficiency), 16-fold more prevalent than for those with less severe FASDs (2.4%). Reported hearing loss was significantly associated with physical features characteristic of FAS.

CONCLUSION: Children with FAS but not other FASDs may be at a particular risk for hearing loss. However, listening difficulties in the absence of hearing loss-presumably related to suprathreshold processing deficits-are prevalent across the entire fetal alcohol spectrum. The nature and impact of both listening difficulties and hearing loss in FASDs warrant further investigation.

Int J Pediatr Otorhinolaryngol. 2019 Oct;125:23-31. doi: 10.1016/j.ijporl.2019.06.015. Epub 2019 Jun 20.

[Enhancing language in children who are deaf/hard-of-hearing using augmentative and alternative communication technology strategies.](#)

[Meinzen-Derr J](#), [Sheldon RM](#), [Henry S](#), [Grether SM](#), [Smith LE](#), [Mays L](#), [Riddle I](#), [Altaye M](#), [Wiley S](#).

BACKGROUND: Despite early identification and intervention, many children who are deaf/hard of hearing (D/HH) demonstrate significant gaps in language development which can directly impact social interactions.

AIMS: The objective of this pilot study was to determine whether integrating augmentative and alternative communication (AAC) core word language strategies into a speech-language therapy program for young children who are D/HH improves spoken language outcomes.

METHODS: Eleven young children, median age 5 years 7 months (range 3y;11 m to 10y;8m) with bilateral hearing loss were enrolled in a single-case experimental design and completed a 24-week intervention that incorporated high-tech AAC strategies into a traditional speech-language therapy model (technology-assisted language intervention or TALi). The goal of the TALi was to improve spoken language development in children who were D/HH. Language samples were collected throughout the study and pragmatic language was assessed pre and post intervention.

RESULTS: At the end of 24 weeks, children demonstrated a significant increase in their mean length of utterance, number of words spoken, and mean turn length according to language samples. Children also made gains in their pragmatic skills pre to post intervention.

CONCLUSIONS: Results of this study suggest that using AAC core word language strategies delivered via iPad technology may support continued and rapid spoken language skill growth among young school-age children who are D/HH. By leveraging AAC technology, we are pioneering a structured and dynamic approach to language learning, building an effective foundation for concepts and grammar for children who are D/HH.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109650. doi: 10.1016/j.ijporl.2019.109650. Epub 2019 Aug 21.

[Vestibular rehabilitation exercises programs to improve the postural control, balance and gait of children with sensorineural hearing loss: A systematic review.](#)

[Melo RS](#), [Lemos A](#), [Paiva GS](#), [Ithamar L](#), [Lima MC](#), [Eickmann SH](#), [Ferraz KM](#), [Belian RB](#).

BACKGROUND: Several studies have demonstrated that children with sensorineural hearing loss (SNHL) exhibit postural instabilities, as well as balance and gait disorders, due to the vestibular dysfunction that they are prone to display as a consequence of inner ear injury. Thus, some experiments have proposed vestibular rehabilitation exercises programs as a treatment to improve these motor skills in children with SNHL.

OBJECTIVE: Assess the evidence quality of the trials that used vestibular rehabilitation exercises programs to improve the postural control, balance and gait of children with SNHL.

METHODS: This is a systematic review that surveyed articles in nine databases, published up to July 4, 2019, in any language, using the following inclusion criteria: (1) Randomized or quasi-randomized controlled trials. (2) Participants of both groups with clinical diagnosis of SNHL, aged up to 12 years old, with no physical problems, cognitive or neurological impairments, except the vestibular dysfunction. (3) Using vestibular rehabilitation exercises programs to improve the following outcomes: postural control, balance and/or gait.

RESULTS: Six experiments, including 153 children, met the inclusion criteria of this systematic review. Two randomized controlled trials (45 children) on the postural control exhibited low evidence quality and four others; three randomized and controlled trials (90 children) on the balance and one quasi-randomized (18 children) on the gait demonstrated very low evidence quality, respectively.

CONCLUSION: There is promising evidence that vestibular rehabilitation exercises programs improve the postural control, balance and gait of children with SNHL. However, due to the methodological limitations of the trials and low quality of current evidence on this topic, the trials results analyzed by this systematic review should be interpreted with caution. Due to the low quality of evidence observed in this review, we suggest that new trials be proposed on this topic, with better methodological quality, to prove the effectiveness of vestibular rehabilitation exercises programs to improve the postural control, balance and gait of children with SNHL.

Braz J Otorhinolaryngol. 2019 Jun 18. pii: S1808-8694(19)30058-8. doi: 10.1016/j.bjorl.2019.05.006. [Epub ahead of print]

Development of auditory and language skills in children using cochlear implants with two signal processing strategies.

Melo TM, Yamaguti EH, Moret ALM, Costa OA, Lopes NBF.

INTRODUCTION: The increase in the spectral information offered by the sound processing strategy HiRes 120 has led to great expectations for the pediatric population. Due to a shorter duration of auditory deprivation and higher neural plasticity, children could benefit more substantially from the spectral information of this sound processing strategy.

OBJECTIVE: To compare auditory and language skills in Brazilian children with cochlear implants using the HiRes and HiRes 120 sound processing strategies.

METHODS: Thirty children, aged 1-3 years, with congenital hearing loss, were divided into two groups, according to the signal processing strategy adjusted at the time of the cochlear implant activation. The assessed children were matched according to chronological age and the time of the cochlear implant use. The auditory and language skills were evaluated longitudinally through the Infant-Toddler Meaningful Auditory Integration Scale and Production Infant Scale Evaluation, carried out before surgery, and 3, 6 and 12 months after device implantation. The Mann-Whitney test was applied for the comparison between the two groups with a 5% significance level.

RESULTS: The findings indicated development of hearing and language skills in the first year of cochlear implant use; however, there was no statistically significant difference in the evolution of such skills due to the adjusted processing strategy in the activation of the cochlear implant electrodes.

CONCLUSION: The development of auditory and language skills in the assessed children was similar during the entire study period, regardless of which signal processing strategy was used.

Am J Audiol. 2019 Sep 13;28(3):597-604. doi: 10.1044/2019_AJA-18-0132. Epub 2019 Jul 24.

Spanish Pediatric Speech Recognition Threshold Test.

Mendel LL, Pousson M, Bass JK, Lunsford RE, McNiece C.

The purpose of this study was to construct a recorded speech recognition threshold (SRT) test for Spanish-speaking children utilizing a picture board and a picture-pointing task.

DESIGN: The Spanish Pediatric Speech Recognition Threshold (SPSRT) test was developed and validated in this study. Test construction steps included (a) stimulus selection, (b) assessment of familiarity, (c) digital recording, (d) creation of pictures that accurately depicted the target word from the stimulus set, and (e) validation of the test and recordings. SRTs were obtained from 24 Spanish-speaking children whose 1st language was Spanish.

RESULTS: Normative data are presented that validate the SPSRT and establish the baseline relationship between the pure-tone average and the SRT obtained with the SPSRT. Results indicated that the SPSRT obtained using this test should be within 2-12 dB of an individual's pure-tone average for Spanish-speaking children with normal hearing and minimal hearing loss.

CONCLUSIONS: The SPSRT was developed and validated as a picture-pointing Spanish SRT test to be used with Spanish-speaking children. The 2-channel recording contains an English translation track, making this test easy to administer and interpret for clinicians without knowledge of Spanish.

Recording Obligatory Cortical Auditory Evoked Potentials in Infants: Quantitative Information on Feasibility and Parent Acceptability.

[Munro KJ](#), [Purdy SC](#), [Uus K](#), [Visram A](#), [Ward R](#), [Bruce IA](#), [Marsden A](#), [Stone MA](#), [Van Dun B](#).

OBJECTIVES: With the advent of newborn hearing screening and early intervention, there is a growing interest in using supra-threshold obligatory cortical auditory evoked potentials (CAEPs) to complement established pediatric clinical test procedures. The aim of this study was to assess the feasibility, and parent acceptability, of recording infant CAEPs.

DESIGN: Typically developing infants ($n = 104$) who had passed newborn hearing screening and whose parents expressed no hearing concerns were recruited. Testing was not possible in 6 infants, leaving 98, age range 5 to 39 weeks (mean age = 21.9, SD = 9.4). Three short duration speech-like stimuli (/m/, /g/, /t/) were presented at 65 dB SPL via a loudspeaker at 0° azimuth. Three criteria were used to assess clinical feasibility: (i) median test duration <30 min, (ii) >90% completion rate in a single test session, and (iii) >90% response detection for each stimulus. We also recorded response amplitude, latency, and CAEP signal to noise ratio. Response amplitudes and residual noise levels were compared for Fpz ($n = 56$) and Cz ($n = 42$) noninverting electrode locations. Parental acceptability was based on an 8-item questionnaire (7-point scale, 1 being best). In addition, we explored the patient experience in semistructured telephone interviews with seven families.

RESULTS: The median time taken to complete 2 runs for 3 stimuli, including preparation, was 27 min (range 17 to 59 min). Of the 104 infants, 98 (94%) were in an appropriate behavioral state for testing. A further 7 became restless during testing and their results were classified as “inconclusive.” In the remaining 91 infants, CAEPs were detected in every case with normal bilateral tympanograms. Detection of CAEPs in response to /m/, /g/, and /t/ in these individuals was 86%, 100%, and 92%, respectively. Residual noise levels and CAEP amplitudes were higher for Cz electrode recordings. Mean scores on the acceptability questionnaire ranged from 1.1 to 2.6. Analysis of interviews indicated that parents found CAEP testing to be a positive experience and recognized the benefit of having an assessment procedure that uses conversational level speech stimuli.

CONCLUSIONS: Test duration, completion rates, and response detection rates met (or were close to) our feasibility targets, and parent acceptability was high. CAEPs have the potential to supplement existing practice in 3- to 9-month olds.

Otolaryngol Clin North Am. 2020 Feb;53(1):87-102. doi: 10.1016/j.otc.2019.09.004. Epub 2019 Oct 31.

Cochlear Implant.

[Naples JG](#), [Ruckenstein MJ](#).

Cochlear implant is the first approved cranial nerve stimulator that works by directly stimulating the cochlear nerve. The medical and societal impact of this revolutionary device cannot be understated. This article reviews the evolving indications for cochlear implant, patient assessment, surgical approach, and outcomes for pediatric and adult cochlear implant that demonstrate its impact. Future concepts in cochlear implant are introduced briefly. This article covers a breadth of information; however, it is not intended to be entirely comprehensive. Rather, it should serve as a foundation for understanding cochlear implant.

J Am Acad Audiol. 2019 Sep;30(8):703-711. doi: 10.3766/jaaa.17126. Epub 2019 Apr 17.

Preimplant Hearing Aid Fittings and Aided Audibility for Pediatric Cochlear Implant Recipients.

[Nickerson A](#), [Davidson LS](#), [Uchanski RM](#).

BACKGROUND: Audibility of speech for children with hearing loss (HL) depends on the degree of HL and the fitting of the hearing aids (HAs) themselves. Many studies on cochlear implant (CI) users have demonstrated that preimplant hearing is associated with postimplant outcomes, but there have been very few reports on the fitting of HAs before surgery.

PURPOSE: The aims of this study were to characterize HA fittings and aided audibility of speech for pediatric HA users with severe to profound HL and to examine the relation between preimplant aided audibility and postimplant speech perception.

RESEARCH DESIGN: A descriptive/observational and correlational study. Audiologic records of pediatric CI participants involved in a larger study examining the effects of early acoustic hearing were analyzed retrospectively; when available, these records included HA verification and speech recognition performance.

STUDY SAMPLE: The CI participants were enrolled in audiology centers and oral schools for the deaf across the United States.

DATA COLLECTION AND ANALYSIS: To determine whether deviations from prescribed DSL target were significantly greater than zero, 95% confidence intervals of the mean deviation were calculated for each frequency (250, 500, 1000, 2000, and 4000 Hz). Correlational analyses were used to examine the relationship between preimplant aided Speech Intelligibility Indices (SIIs) and postimplant speech perception in noise. Correlational analyses were also used to explore the relationship between preimplant aided SIIs and demographic data. T-tests were used to compare preimplant-aided SIIs of HAs of listeners who later became users of either sequential CIs, simultaneous CIs, or bimodal devices.

RESULTS: Preimplant fittings of HAs were generally very close to prescriptive targets, except at 4000 Hz for those HAs with active frequency-lowering processing, and preimplant SIIs, albeit low, were correlated with postimplant speech recognition performance in noise. These results suggest that aided audibility should be maximized throughout the HA trial for later speech recognition purposes.

CONCLUSIONS: It is recommended that HA fittings be optimized to support speech audibility even when considering implantation. In addition to the age at which HA use begins, the aided audibility itself is important in determining CI candidacy and decisions regarding bimodal HA use.

J Clin Oncol. 2019 Jun 20;37(18):1566-1575. doi: 10.1200/JCO.18.01358. Epub 2019 May 2.

Cognitive Implications of Ototoxicity in Pediatric Patients With Embryonal Brain Tumors.

Olivier TW, Bass JK, Ashford JM, Beaulieu R, Scott SM, Schreiber JE, Palmer S, Mabbott DJ, Swain MA, Bonner M, Boyle R, Chapeiski ML, Evankovich KD, Armstrong CL, Knight SJ, Wu S, Onar-Thomas A, Gajjar A, Conklin HM.

PURPOSE: Sensorineural hearing loss (SNHL) is associated with intellectual and academic declines in children treated for embryonal brain tumors. This study expands upon existing research by examining core neurocognitive processes that may result in reading difficulties in children with treatment-related ototoxicity.

PATIENTS AND METHODS: Prospectively gathered, serial, neuropsychological and audiology data for 260 children and young adults age 3 to 21 years (mean, 9.15 years) enrolled in a multisite research and treatment protocol, which included surgery, risk-adapted craniospinal irradiation (average risk, $n = 186$; high risk, $n = 74$), and chemotherapy, were analyzed using linear mixed models. Participants were assessed at baseline and up to 5 years after diagnosis and grouped according to degree of SNHL. Included were 196 children with intact hearing or mild to moderate SNHL (Chang grade 0, 1a, 1b, or 2a) and 64 children with severe SNHL (Chang grade 2b or greater). Performance on eight neurocognitive variables targeting reading outcomes (eg, phonemics, fluency, comprehension) and contributory cognitive processes (eg, working memory, processing speed) was analyzed.

RESULTS: Participants with severe SNHL performed significantly worse on all variables compared with children with normal or mild to moderate SNHL ($P \leq .05$), except for tasks assessing awareness of sounds and working memory. Controlling for age at diagnosis and risk-adapted craniospinal irradiation dose, performance on the following four variables remained significantly lower for children with severe SNHL: phonemic skills, phonetic decoding, reading comprehension, and speed of information processing ($P \leq .05$).

CONCLUSION: Children with severe SNHL exhibit greater reading difficulties over time. Specifically, they seem to struggle most with phonological skills and processing speed, which affect higher level skills such as reading comprehension.

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Determining concordance and cost impact of otoacoustic emission and automated auditory brainstem response in newborn hearing screening in a tertiary hospital.

Ong KMC, Rivera AS, Chan AL, Chiong CM.

OBJECTIVE: This study compared otoacoustic emission (OAE) and automated auditory brainstem response (AABR) in terms of concordance and cost impact for newborn hearing screening (NBHS) in the Philippine setting.

METHODS: This was a prospective observational study to assess concordance between OAE and AABR involving 253 infants. Each infant underwent OAE and AABR testing. Infants who passed both tests were not required to follow up for additional testing. Infants who failed in any test were scheduled for repeat screening and diagnostic ABR after 1 month. Concordance was computed using B-statistic.

FOR COST ANALYSIS: 4 scenarios were compared to 1-step both tests scenario: (1) OAE alone, (2) AABR alone, (3) 2-step OAE, and (4) 2-step AABR in terms of number of infants with hearing loss (HL) detected, cost of diagnosis, and economic loss from lack of treatment.

RESULTS: There was high concordance between OAE and AABR (B-statistic=0.8). AABR had a higher refer rate (18.58%) than OAE (10.27%) but higher number of detected babies with HL. Cost analysis favored an AABR alone scenario while the 2-step OAE protocol fared poorly.

CONCLUSION: A change from 2-step OAE to AABR alone is worth considering in our institution.

First Pediatric Experience With a Novel, Adhesive Adapter Retained, Bone Conduction Hearing Aid System.

[Osborne MS](#), [Child-Hymas A](#), [Gill J](#), [Lloyd MS](#), [McDermott AL](#).

OBJECTIVES: To assess the audiological outcomes, practicalities, and impact on quality of life of a new, nonimplantable, adhesive retained bone conduction hearing aid in children.

STUDY DESIGN: A prospective, single-subject repeat measure, cohort study.

SETTING: Community and in pediatric assessment center.

PATIENTS: Twenty-one children aged between 5 and 15 years with a conductive hearing loss of ≥ 25 dB HL in the better hearing ear.

INTERVENTION: Audiological comparisons were made using pure-tone thresholds; unaided, with a softband aid, and with the new adhesive retained bone conducting system.

MAIN OUTCOME MEASURES: Comparison of hearing threshold levels. Data analysis via paired t-testing, significance set at p value < 0.01 . Quality of life was assessed via the Glasgow Children's Benefit Inventory and a 10cm linear analogue scale. A hearing aid review questionnaire provided insight into practical use.

RESULTS: Statistically significant improvement in thresholds of 7.3 dB HL ($p = 0.0001$) was demonstrated with the adhesive system as compared with softband aids. After 4 weeks of usage, the mean hearing thresholds for the adhesive hearing system improved from 55 dB HL ± 2.4 to 31 dB HL ± 7.9 in unaided and aided conditions. Improvements in QOL were demonstrated with LAS and GCBI. Four children reported mild skin reactions. Eighty-six percent reported improved self-confidence.

CONCLUSION: The adhesive aid produces comparable audiological results to the commercial softband hearing aids. It provides an excellent alternative in the treatment of conductive hearing loss without the possible complications and costs of a surgical intervention. Furthermore, it preserves skin envelope over the mastoid for those who wish to proceed with an autologous pinna reconstruction in the future.

Neonatal screening for congenital CMV infection stresses the importance of maternal nonprimary infection even in an area where prenatal serology testing is common.

[Papaevangelou V](#), [Christoni Z](#), [Vliora C](#), [Kottaridi C](#), [Fotiou A](#), [Malamitsi-Puchner A](#), [Mentis A](#), [Karakitsos P](#), [Syggelou A](#).

AIM AND METHODS: Dried blood spots from 2149 newborns were examined to diagnose congenital cytomegalovirus (cCMV).

RESULTS: Prenatal CMV-IgG antibodies had been measured during prenatal care in 1287 (60.3%) of mothers and 980 (76.1%) of them were found seropositive. cCMV incidence was 0.47%. All newborns were asymptomatic; 9/10 were born post nonprimary maternal infection; two developed sensorineural hearing loss.

CONCLUSIONS: In a country where prenatal CMV testing is common and therefore a false sense of control might prevail, nonprimary maternal infection should not be overlooked. Indeed, women of childbearing age should be educated on CMV prevention measures irrespectively to their serostatus.

Hearing of neonates without risk indicators for hearing loss and use of antimalarial drugs during pregnancy: a historical cohort study in the Northern Region of Brazil.

[Patatt FSA](#), [Sampaio ALL](#), [Tauil PL](#), [Oliveira CACP](#).

INTRODUCTION: Studies have demonstrated the ototoxic effects of antimalarial drugs in individuals who receive these drugs, but little is known about their toxicity in the neonatal auditory system when the mothers receive the drug during pregnancy.

OBJECTIVE: To verify the incidence of hearing loss in neonates who have no other associated risk indicators, born to mothers treated for malaria during pregnancy.

METHODS: A retrospective, quantitative cohort study was developed at Hospital de Base Dr. Ary Pinheiro and Clínica Limiar, both located in the municipality of Porto Velho (Rondônia). The sample consisted of 527 newborns divided into two groups: exposed to antimalarials drugs during pregnancy group ($n = 32$) and non-exposed group

(n=495). Data collection took place from September 2014 to December 2015, through an interview with the mothers and/or guardians of the newborn, through the newborns' and the mothers' records, and the neonatal hearing screening database of the above-mentioned institutions.

RESULTS: All the neonates in the exposed group, assessed through the recording of transient otoacoustic emissions associated with the automated brainstem auditory evoked potential test, underwent neonatal hearing screening in the first examination. Among the newborns in the non-exposed group, 30 showed failure and were retested. Of these, one continued to fail and was referred for diagnosis, in whom the results showed to be within the normal range. Among the neonates of the exposed group, infection with *Plasmodium vivax* was the most frequent, and was similarly distributed among the gestational trimesters, and chloroquine was the most commonly used antimalarial drug treatment more often given during the third trimester; these findings did not show any influence on the audiological findings of the studied neonates.

CONCLUSION: The present study did not identify any cases of hearing loss in neonates born to mothers who used antimalarial drugs during gestation.

BMC Infect Dis. 2019 Jul 22;19(1):652. doi: 10.1186/s12879-019-4296-5.

Diagnosis of congenital CMV infection via DBS samples testing and neonatal hearing screening: an observational study in Italy.

[Pellegrinelli L](#), [Galli C](#), [Primache V](#), [Alde' M](#), [Fagnani E](#), [Di Berardino F](#), [Zanetti D](#), [Pariani E](#), [Ambrosetti U](#), [Binda S](#).

BACKGROUND: Congenital Cytomegalovirus (cCMV) is the most common cause of non-genetic hearing loss in childhood. A newborn hearing screening program (NHSP) is currently running in Italy, but no universal cCMV nor statewide hearing-targeted CMV screening programs have been implemented yet. This observational monocentric study was aimed at estimating the rate of cCMV infections identified by CMV-DNA analysis on Dried Blood Spots (DBS) samples in deaf children identified via NHSP in Northern Italy in the period spanning from 2014 to 2018.

METHODS: Children with a confirmed diagnosis of deafness and investigated for CMV-DNA by nucleic acid extraction and in-house polymerase-chain reaction (PCR) on stored newborns screening cards (DBS-test) were included in this study. Deafness was defined by a hearing threshold ≥ 20 decibel (dB HL) by Auditory Brainstem Responses (ABR); all investigated DBS samples were collected within 3 days of life.

RESULTS: Overall, 82 children were included (median age: 3.4 months; lower-upper quartiles: 2-5.3 months; males: 60.9%). Most of them (70.7%) presented bilateral hearing loss with a symmetrical pattern in 79.3% of the cases. ABR thresholds were ≥ 70 dB HL (severe/profound deafness) in 46.5% of children. Among all tested children, 6.1% resulted positive for cCMV. The rate of severe/profound deafness was statistically higher in children with cCMV infection.

CONCLUSIONS: The addition of DBS-test to the NHSP allowed the identification, in their first months of life, of a cCMV infection in 6.1% of children who had failed NHS. The introduction of a targeted CMV screening strategy could help clinicians in the differential diagnosis and in the babies' management. DBS samples can be considered a "universal newborns biobank": their storage site and duration should be the subject of political decision-making.

Bull World Health Organ. 2019 Oct 1;97(10):663-671. doi: 10.2471/BLT.18.220939. Epub 2019 Sep 3.

Pilot implementation of newborn hearing screening programme at four hospitals in southern Thailand.

[Pitathawatchai P](#), [Khaimook W](#), [Kirtsreesakul V](#).

OBJECTIVE: To determine the effectiveness and benefit of a universal newborn hearing screening programme at four different hospitals in southern Thailand, between January and July 2017.

METHODS: One screener per hospital recorded demographic data of all newborns and their exposure to risk of hearing loss, and evaluated their hearing by transient otoacoustic emission technology. Those who demonstrated bilateral moderate to profound hearing loss at both a first and second screening were referred for diagnostic assessment. Those with confirmed hearing loss received treatment and regular follow-up appointments, and their speech development was assessed at 1 year of age. We determined effectiveness by comparing our achieved coverage and proportion of follow-up and referrals with benchmarks set by the American Academy of Pediatrics ($\geq 95\%$, $\geq 95\%$ and $\leq 4\%$, respectively), and determined benefit by calculating the composite language scores of hearing-impaired infants who received early intervention.

FINDINGS: We screened 6140 eligible newborns, and achieved a screening coverage of 95.4% (5859/6140), lost 25.7% (63/245) and 22.0% (9/41) to follow-up at the second screening and diagnostic assessment stages, respectively, and obtained an overall proportion of referrals of 0.7% (41/6140). Twelve infants were confirmed as having hearing loss and received early intervention; nine (75%) demonstrated normal speech development by their first birthday. Our universal hearing screening yielded a prevalence of sensorineural hearing loss of less than 0.1% (3/6140).

CONCLUSION: Although ineffective by American Academy of Pediatrics standards, we demonstrated the benefit of early intervention in infants diagnosed with hearing loss.

The Burden of Congenital Cytomegalovirus Infection: A Prospective Cohort Study of 20 000 Infants in Finland.

[Puhakka L](#), [Lappalainen M](#), [Lönnqvist T](#), [Niemensivu R](#), [Lindahl P](#), [Nieminen T](#), [Seuri R](#), [Nupponen I](#), [Pati S](#), [Boppa S](#), [Saxen H](#).

BACKGROUND: Congenital cytomegalovirus (cCMV) infection is the most common congenital infection and causes significant morbidity. This study was undertaken to evaluate the benefits of screening newborns for cCMV and to understand the cCMV disease burden in Finland.

METHODS: Infants born in Helsinki area hospitals were screened for CMV by testing their saliva with a real-time polymerase chain reaction assay. The CMV-positive infants and matched controls were monitored to determine their neurodevelopmental, audiological, and ophthalmological outcomes at 18 months of age. Griffiths Mental Development Scales, otoacoustic emission and sound field audiometry, and ophthalmologic examination were performed.

RESULTS: Of the 19868 infants screened, 40 had confirmed cCMV infection (prevalence, 2 in 1000 [95% confidence interval, 1.4-2.6 in 1000]). Four (10%) infants had symptomatic cCMV. Griffiths general quotients did not differ significantly between the CMV-positive (mean, 101.0) and control (mean, 101.6) infants ($P = .557$), nor did quotients for any of the Griffiths subscales (locomotion, personal-social, hearing and language, eye and hand, performance) ($P = .173-.721$). Four of 54 CMV-positive ears and 6 of 80 CMV-negative ears failed otoacoustic emission testing ($P = 1.000$). The mean minimal response levels over the frequencies 500 Hz to 4 kHz in the sound field audiometry did not differ between CMV-positive (mean, 34.31-dB hearing level) and control (mean, 32.73-dB hearing level) infants ($P = .338$). No CMV-related ophthalmologic findings were observed.

CONCLUSIONS: The prevalence of cCMV was low, and outcomes at 18 months of age did not differ between the infected infants and healthy control infants. With such a low burden in Finland, universal newborn screening for cCMV seems unwarranted.

Hearing-impaired listeners show increased audiovisual benefit when listening to speech in noise.

[Puschmann S](#), [Daeglau M](#), [Stropahl M](#), [Mirkovic B](#), [Rosemann S](#), [Thiel CM](#), [Debener S](#).

Recent studies provide evidence for changes in audiovisual perception as well as for adaptive cross-modal auditory cortex plasticity in older individuals with high-frequency hearing impairments (presbycusis). We here investigated whether these changes facilitate the use of visual information, leading to an increased audiovisual benefit of hearing-impaired individuals when listening to speech in noise. We used a naturalistic design in which older participants with a varying degree of high-frequency hearing loss attended to running auditory or audiovisual speech in noise and detected rare target words. Passages containing only visual speech served as a control condition. Simultaneously acquired scalp electroencephalography (EEG) data were used to study cortical speech tracking. Target word detection accuracy was significantly increased in the audiovisual as compared to the auditory listening condition. The degree of this audiovisual enhancement was positively related to individual high-frequency hearing loss and subjectively reported listening effort in challenging daily life situations, which served as a subjective marker of hearing problems. On the neural level, the early cortical tracking of the speech envelope was enhanced in the audiovisual condition. Similar to the behavioral findings, individual differences in the magnitude of the enhancement were positively associated with listening effort ratings. Our results therefore suggest that hearing-impaired older individuals make increased use of congruent visual information to compensate for the degraded auditory input.

Genetic testing for congenital non-syndromic sensorineural hearing loss.

[Raymond M](#), [Walker E](#), [Dave I](#), [Dedhia K](#).

INTRODUCTION: Approximately 60% of congenital pediatric hearing loss is of genetic etiology. To evaluate non-syndromic sensorineural hearing loss (NSSNHL), guidelines emphasize the use of comprehensive genetic testing (CGT) with next generation sequencing (NGS), yet these tests have limited accessibility, and potential CGT results may not be well understood. Thus, our objective was to analyze genetic testing practices and results for pediatric patients with NSSNHL.

METHODS: This was a retrospective chart review of pediatric patients (<18 years) diagnosed with NSSNHL from 2014 to 2017 at a tertiary pediatric hospital. Demographics, clinical data, CGT results, genetic testing practices and referral patterns were recorded and descriptively analyzed. Logistic regression models identified patient characteristics associated with pathogenic variants (PV) and variants of unknown significance (VOUS).

RESULTS: 430 patients with congenital NSSNHL were included in the study. Genetic testing was ordered for 28% (n=122) and resulted for 16% (n=68). Most of the ordered tests (89%, n=109) were the CGT panel. A majority (62%, n=97) of the time in which genetic testing was not ordered, a referral for genetics consultation was placed. Amongst those with CGT results, a definitive genetic etiology was identified in 25% (n=13), with less than half due to variants of GJB2/6. At least one PV was identified for 33% (n=18), while at least one VOUS for 93% (n=51). There were no significant differences in PV presence or number of VOUS across any characteristic except race. When compared to Caucasians, African Americans had significantly higher rates of VOUS with a rate ratio and 95% CI of 1.61 [1.11-2.34], $p=0.01$, and Asians trended towards higher rates (1.96 [0.95-4.05], $p=0.06$).

CONCLUSIONS: CGT is of high utility in the identification of relevant genetic variants and definitive genetic etiologies for pediatric patients with NSSNHL. Though guidelines recommend the early use of CGT, there are many barriers to appropriate testing and counseling, leading to low rates of CGT use at this single institution.

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[***Vestibular Screening in Pediatric Patients with Otitis Media.***](#)

[Rehagen SK](#), [Valente M](#), [Lieu JEC](#).

BACKGROUND: Otitis media with effusion (OME) is a common cause of vestibular disturbances in children. However, young children often lack the language to express their symptoms, and it is uncommon to screen children for vestibular impairments.

PURPOSE: The purpose of this study was to develop a screening protocol for children presenting with OME to determine if diagnostic vestibular testing is necessary.

RESEARCH DESIGN: Children with normal hearing (NH), sensorineural hearing loss (SNHL), and conductive hearing loss (CHL) due to OME participated in a vestibular screening.

STUDY SAMPLE: There were 30 participants, ages four to eight years, ranging from 48 to 101 months included in the study: 10 with NH, 11 with CHL due to OME, and 9 with SNHL.

DATA COLLECTION AND ANALYSIS: The vestibular screening consists of patient and parent questionnaires and a functional evaluation. The tests examined coordination, balance, oculomotor function, and nystagmus.

RESULTS: Those with CHL were significantly more likely to display abnormal smoothness of pursuit, as measured with observations for rapid tracking, absence of or delayed saccades, and overshoot, than those with NH or SNHL ($p = 0.012$). Parents of children with CHL due to OME were more likely to report their child experiencing middle ear pressure than the parents of children with NH or SNHL ($p = 0.010$). In addition, children with CHL were less likely to report hearing loss than those with NH or SNHL. Parent and patient report were not found to be reliable indicators of vestibular disturbances.

CONCLUSIONS: This pilot study suggests that children with CHL due to OME present with more oculomotor abnormalities than their peers with NH. Further research is necessary to determine validity and reliability of the findings for this present study.

J Pediatr Health Care. 2019 Nov - Dec;33(6):694-701. doi: 10.1016/j.pedhc.2019.06.001. Epub 2019 Jul 31.

[***Identification of Waardenburg Syndrome and the Management of Hearing Loss and Associated Sequelae: A Review for the Pediatric Nurse Practitioner.***](#)

[Ringer J](#).

Waardenburg syndrome (WS) is a rare genetic disorder that is further divided into four subtypes with distinguishing clinical manifestations, categorized by phenotypic variations based on activation or deactivation of six specific gene types. The criteria for clinical diagnosis are established based on these phenotypic variants. While key clinical features may cause suspicion of WS, genetic testing confirms the diagnosis. Pigmentary defects are one of the hallmark features of WS while some individuals may exhibit sensorineural hearing loss, which can be progressive. Audiological treatment is essential to mitigate hearing loss and to minimize speech and language deficits as well as behavior and socioemotional development. Associated complications include musculoskeletal abnormalities and Hirschsprung disease. This article aims to discuss the role of the pediatric nurse practitioner in the early identification, diagnosis, treatment, and long-term management of affected children in the primary care setting.

J Perinatol. 2019 Oct 1. doi: 10.1038/s41372-019-0501-z. [Epub ahead of print]

[***Evaluation of clinically asymptomatic high risk infants with congenital cytomegalovirus infection.***](#)

[Ronchi A](#), [Zeray F](#), [Lee LE](#), [Owen KE](#), [Shoup AG](#), [Garcia F](#), [Vazquez LN](#), [Cantey JB](#), [Varghese S](#), [Pugni L](#), [Mosca F](#), [Sánchez PJ](#).

OBJECTIVE: To determine the frequency of abnormal findings on evaluation of neonates with congenital CMV infection who have a normal physical examination **STUDY DESIGN:** Retrospective, 2-center study (1996-2017)

that reviewed results of complete blood cell count and platelets, serum alanine aminotransferase (ALT) and bilirubin concentrations, eye examination, cranial ultrasonography or other neuroimaging, and brainstem evoked responses performed on neonates with congenital CMV infection and a normal physical examination RESULTS: Of 34 infants with congenital CMV infection and a normal physical examination, 56% (19/34) had ≥ 1 abnormality: 39%, elevated ALT concentration; 45%, abnormal neuroimaging (five, lenticulostriate vasculopathy; six, intraventricular hemorrhage; four, calcifications); 12%, anemia; 16%, thrombocytopenia; and 3%, chorioretinitis. Seven (21%) infants had sensorineural hearing loss, and 18 infants received antiviral therapy.

CONCLUSION: Some infants with congenital CMV infection and a normal physical examination had abnormalities on laboratory or neuroimaging evaluation, which in some cases prompted antiviral treatment.

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Quality of Life in Children with Sensorineural Hearing Loss.

Ronner EA, Benchetrit L, Levesque P, Basonbul RA, Cohen MS.

OBJECTIVE: To assess quality of life (QOL) in pediatric patients with sensorineural hearing loss (SNHL) with the Pediatric Quality of Life Inventory 4.0 (PedsQL 4.0) and the Hearing Environments and Reflection on Quality of Life 26 (HEAR-QL-26) and HEAR-QL-28 surveys.

STUDY DESIGN: Prospective longitudinal study.

SETTING: Tertiary care center.

SUBJECTS AND METHODS: Surveys were administered to patients with SNHL (ages 2-18 years) from July 2016 to December 2018 at a multidisciplinary hearing loss clinic. Patients aged >7 years completed the HEAR-QL-26, HEAR-QL-28, and PedsQL 4.0 self-report tool, while parents completed the PedsQL 4.0 parent proxy report for children aged ≤ 7 years. Previously published data from children with normal hearing were used for controls. The independent t test was used for analysis.

RESULTS: In our cohort of 100 patients, the mean age was 7.7 years (SD, 4.5): 62 participants had bilateral SNHL; 63 had mild to moderate SNHL; and 37 had severe to profound SNHL. Sixty-eight patients used a hearing device. Mean (SD) total survey scores for the PedsQL 4.0 (ages 2-7 and 8-18 years), HEAR-QL-26 (ages 7-12 years), and HEAR-QL-28 (ages 13-18 years) were 83.9 (14.0), 79.2 (11.1), 81.2 (9.8), and 77.5 (11.3), respectively. Mean QOL scores for patients with SNHL were significantly lower than those for controls on the basis of previously published normative data ($P < .0001$). There was no significant difference in QOL between children with unilateral and bilateral SNHL or between children with SNHL who did and did not require a hearing device. Low statistical power due to small subgroup sizes limited our analysis.

CONCLUSION: It is feasible to collect QOL data from children with SNHL in a hearing loss clinic. Children with SNHL had significantly lower scores on validated QOL instruments when compared with peers with normal hearing.

JAMA Otolaryngol Head Neck Surg. 2019 May 1;145(5):431-443. doi: 10.1001/jamaoto.2019.0121.

Assessment of the Clinical Benefit of Imaging in Children With Unilateral Sensorineural Hearing Loss: A Systematic Review and Meta-analysis.

Ropers FG, Pham ENB, Kant SG, Rotteveel LJC, Rings EHHM, Verbist BM, Dekkers OM.

IMPORTANCE: Imaging used to determine the cause of unilateral sensorineural hearing loss (USNHL) in children is often justified by the high likelihood of detecting abnormalities, which implies that these abnormalities are associated with hearing loss and that imaging has a positive contribution to patient outcome or well-being by providing information on the prognosis, hereditary factors, or cause of hearing loss.

OBJECTIVES: To evaluate the diagnostic yield of computed tomography (CT) and magnetic resonance imaging (MRI) in children with isolated unexplained USNHL and investigate the clinical relevance of these findings.

EVIDENCE REVIEW: Cochrane Library, Embase, PubMed, and Web of Science databases were searched for articles published from 1978 to 2017 on studies of children with USNHL who underwent CT and/or MRI of the temporal bone. Two authors (F.G.R. and E.N.B.P.) independently extracted information on population characteristics, imaging modality, and the prevalence of abnormalities and assessed the studies for risk of bias. Eligibility criteria included studies with 20 or more patients with USNHL who had CT and/or MRI scans, a population younger than 18 years, and those published in English.

MAIN OUTCOMES AND MEASURES: The pooled prevalence with 95% CI of inner ear abnormalities grouped according to finding and imaging modality.

FINDINGS: Of 1562 studies, 18 were included with a total of 1504 participants included in the analysis. Fifteen studies were consecutive case studies and 3 were retrospective cohort studies. The pooled diagnostic yield for pathophysiologic relevant findings in patients with unexplained USNHL was 37% for CT (95% CI, 25%-48%) and

35% for MRI (95% CI, 22%-49%). Cochleovestibular abnormalities were found with a pooled frequency of 19% for CT (95% CI, 14%-25%) and 16% for MRI (95% CI, 7%-25%). Cochlear nerve deficiency and associated cochlear aperture stenosis had a pooled frequency of 16% for MRI (95% CI, 3%-29%) and 44% for CT (95% CI, 36%-53%), respectively. Enlarged vestibular aqueduct (EVA) was detected with a pooled frequency of 7% for CT and 12% for MRI in children with USNHL.

CONCLUSIONS AND RELEVANCE: Imaging provided insight into the cause of hearing loss in a pooled frequency of about 35% to 37% in children with isolated unexplained USNHL. However, none of these findings had therapeutic consequences, and imaging provided information on prognosis and hereditary factors only in a small proportion of children, namely those with EVA. Thus, there is currently no convincing evidence supporting a strong recommendation for imaging in children who present with USNHL. The advantages of imaging should be carefully balanced against the drawbacks during shared decision making.

Int J Pediatr Otorhinolaryngol. 2019 Sep 20;128:109687. doi: 10.1016/j.ijporl.2019.109687. [Epub ahead of print]

[The Professional's experience with causes of delay in the diagnosis and management of children with a congenital hearing loss in Libya.](#)

[Sambah I, Zhao F, El-Lishani R.](#)

The aim of this study was to collect and interpret narrative and observational data from Audiologists and ENT doctors' experiences of delays in the identification and management of congenital hearing loss (CHL) in Libya. This qualitative study sought to explore and understand the reasons behind the delay. Participants were three Audiological Physicians and five Otolaryngologists (ENT) working in public hospitals in four large cities in Libya. They were interviewed to explore the causes of such delays and themes were generated from their experiences. All participants revealed that the main causes might be associated with limited facilities and availability of audiology services, lack of awareness and knowledge of the magnitude of the issue and the importance of early detection and intervention for CHL in Libya. In contrast to other developing countries, the financial situation and poverty were not considered to be the main cause in Libya. Furthermore, socioeconomic status of the children's families appears relevant.

Int J Pediatr Otorhinolaryngol. 2019 Aug;123:123-127. doi: 10.1016/j.ijporl.2019.05.001. Epub 2019 May 8.

[Variations of the vascular canals in the cochlear implant candidates.](#)

[Sarioglu FC, Pekcevik Y, Guleryuz H, Olgun Y, Guneri EA.](#)

OBJECTIVE: To evaluate the incidence of vascular canal variations in the pediatric cochlear implant (CI) candidates.

METHODS: We retrospectively reviewed temporal bone computed tomography (CT) images of the CI candidates between November 2013 and November 2018. The presence of high riding jugular bulb, dehiscent jugular bulb, jugular bulb diverticulum, bulging of sigmoid sinus, mastoid emissary vein (MEV), carotid canal dehiscence, and aberrant internal carotid canal were evaluated. Findings were compared with a control group of normal-hearing subjects.

RESULTS: Temporal CT images of 118 CI candidates and 119 control group participants were evaluated. The vascular canal anomalies were found in 88 (37.3%) temporal bones of the CI candidates and 49 (20.6%) of the control group ($p < 0.001$). In 236 temporal CT scans of the CI candidates and 238 temporal CT scans of the control group, we found MEV in 19.1% and 6.3%, high riding jugular bulb in 11.4% and 10.5%, dehiscent jugular bulb in 2.1% and 1.3%, jugular bulb diverticulum in 6.4% and 1.7%, bulging sigmoid sinus in 11.4% and 4.2%, carotid canal dehiscence in 0.8% and 1.3%, and aberrant internal carotid canal in 0 and 0.8%, respectively. Jugular bulb diverticulum ($p = 0.01$), bulging of the sigmoid sinus ($p = 0.003$), and MEV ($p < 0.001$) were more frequent in the CI candidates.

CONCLUSION: Vascular canal variations are more common in the CI candidates and should be evaluated before CI surgery.

Indian J Otolaryngol Head Neck Surg. 2019 Sep;71(3):401-405. doi: 10.1007/s12070-017-1117-2. Epub 2017 Apr 13.

[Clinical Experience on Hearing Screening in Twins and Triplets: A Retrospective Study.](#)

[Sasireka BI, Jaya V, Vignesh SS, Muthukumar R.](#)

Twin or multiple pregnancies often have neonatal complications. Common complications include low birth weight, respiratory distress, neonatal intensive care unit (NICU) admission (> 5 days) and low APGAR score especially on second twin. (1) To compare referral rates of newborn hearing screening in twins and triplets between risk and non-risk babies for hearing loss, (2) to determine the effects of birth order on referral rates in twins and

triplets. A retrospective study was carried out from the case records of the neonates enrolled in the referral based NICU hearing screening program during April 2013 to December 2014 at Institute of Obstetrics and Gynecology, Chennai. 1405 neonates (723 males and 682 females) in the age range of 3-28 days were screened during this period among which 76 were twins (38 pairs) and 9 were triplets (3 sets). We classified them further into non risk and risk babies among twins according to the Joint Committee for Infant Hearing (JCIH) recommendations. A dual step hearing screening protocol was used to screen the neonates. They were initially tested with distortion product otoacoustic emission (DPOAE) then with automated auditory brainstem response (AABR) before discharged from the hospital. Out of 76 (100%) twin babies 43 (56.58%) babies had one or more risk factors for hearing loss. Most common risk factors were low birth weight, premature birth, mechanical ventilation >5 days, hyperbilirubinemia, infections, ototoxic medications and family history of hearing loss. Each risk factor was observed in equal proportion in 1st born and 2nd born twins. 12 (15.76%) risk babies and 11 (14.47%) non risk babies failed in DPOAE testing. Out of 9 triplets babies (3 set) 3 babies had risk factors for hearing loss. One baby in 2nd born babies group failed AABR screening. Referral rates were slightly higher in 2nd born twins when compared to 1st born twins in DPOAE testing. Chi square test did not reveal any significant association between referral rates with risk factors and birth order ($p > 0.05$). The referral rates in twins are higher than the total population screened. There is some influence of birth order on the referral rates in twins. It needs to be further investigated on larger population.

Indian J Otolaryngol Head Neck Surg. 2019 Nov;71(Suppl 2):1383-1390. doi: 10.1007/s12070-018-1454-9. Epub 2018 Jul 16.

[Screening of Newborn Hearing at a Tertiary Care Hospital in South India.](#)

[Satish HS](#), [Anil Kumar R](#), [Viswanatha B](#).

Hearing loss can have a devastating impact on the cognitive development and psychological well-being of children and their families. Newborn hearing screening should be given special attention especially in a country like ours where the burden of this disability is heavy. Screening all newborns irrespective of risk factors helps in better detection, and hence further management can be initiated at appropriate time. To implement newborn hearing screening at Vanivilas hospital and to estimate the incidence of hearing loss among the high risk groups. To create awareness about the need for detecting childhood deafness among parents and general population. To develop a centre of excellence for evaluation, intervention and rehabilitation for hearing impaired in a tertiary care hospital. Prospective study. All neonates born in Vanivilas Hospital attached to Bangalore Medical College and Research Institute underwent hearing screening using four stage protocols with otoacoustic emission (OAE) tests and final confirmation with Brainstem evoked response audiometry (BERA) tests. May 2015-May 2017. Number of newborns screened were 26,487, and 19 (0.717/1000) newborns were detected to have hearing impairment. The incidence of hearing loss among high risk group was 0.188/1000, and among the non risk group was 0.528/1000. Newborn hearing screening must be made mandatory and multi-staged protocol based screening for hearing loss should be implemented. This will make newborn screening programme more efficient and also will help in initiating treatment at an early stage so that further damage can be prevented.

© Association of Otolaryngologists of India 2018.

Otol Neurotol. 2019 Oct;40(9):1148-1152. doi: 10.1097/MAO.0000000000002342.

[Factors Influencing Pediatric Cochlear Implant Outcomes: Carolina Sibling Study.](#)

[Selleck AM](#), [Park LR](#), [Brown KD](#).

OBJECTIVE: To determine the correlation between speech perception performance between pediatric sibling pairs with severe to profound sensory hearing loss receiving cochlear implants, and in cases of discordance in performance, determine which variables negatively impacted performance.

STUDY DESIGN: Retrospective chart review.

SETTING: Tertiary academic referral center.

SUBJECTS: Eighty-nine pediatric subjects, 43 sibling groups, under the age of 18 with severe to profound sensory hearing loss who received a cochlear implant with a sibling who also received a cochlear implant.

INTERVENTION: Cochlear implantation.

MAIN OUTCOME MEASURES: Speech perception scores, consonant-nucleus- consonant score.

RESULTS: A statistically significant correlation was found between speech perception performance of pediatric siblings. Depth of insertion is positively correlated with better speech perception in siblings with discordant results. There was no significant relationship with either electrode type, unilateral/bilateral status, or age of implantation.

CONCLUSION: Pediatric siblings have a high correlation in speech perception outcomes following cochlear implantation, suggesting family environment plays a strong role. In circumstances in which outcomes between siblings are substantially different, greater depth of implant insertion is correlated with better consonant-nucleus-consonant word scores.

Cleft Palate Craniofac J. 2019 Jul;56(6):720-728. doi: 10.1177/1055665618809228. Epub 2018 Nov 11.

[The Impact of Timing of Tympanostomy Tube Placement on Sequelae in Children With Cleft Palate.](#)

[Shaffer AD](#), [Ford MD](#), [Choi SS](#), [Jabbour N](#).

OBJECTIVE: To describe the impact of timing of tympanostomy tube insertion on the number of tubes received and complications in children with routine tube placement.

DESIGN: Retrospective case series.

SETTING: Tertiary care children's hospital.

PARTICIPANTS: Records from a consecutive sample of 401 children with cleft palate were reviewed. Sixty-five patients with isolated cleft palate and 82 patients with cleft lip and palate had follow-up until 5 years of age and were included.

INTERVENTIONS: Tympanostomy tubes.

MAIN OUTCOME MEASURE(S): Number of tubes received and tube-related complications. The hypothesis was formulated prior to data collection.

RESULTS: Males comprised 55.8% of included patients, and tubes were placed in 98.6% of patients at a median age of 6.5 months. Effusion was documented at first tube placement for 96.5% of patients. Most (67.4%) patients required replacement of tubes, and 10.6% required long-term tubes. Complications included otorrhea (71.0%), myringosclerosis (35.2%), granulation (22.8%), perforation (17.9%), retained tubes (5.5%), and cholesteatoma (1.4%). Cleft lip and palate ($P < .001$) and otorrhea ($P = .023$) were associated with tube placement before palatoplasty. Patients with tube placement before palatoplasty ($P = .033$), genetic disorders ($P = .007$), failed newborn hearing screen ($P = .012$), otorrhea ($P < .001$), and granulation ($P < .001$) received more tubes.

CONCLUSIONS: Nearly universal effusion in patients with cleft palate supports the need for routine tube placement. The potential for otorrhea and requiring more tubes should be weighed against the risks associated with prolonged effusion when considering tube placement before palatoplasty.

Otol Neurotol. 2019 Jun;40(5):e518-e526. doi: 10.1097/MAO.0000000000002227.

[Electrical Impedance as a Biomarker for Inner Ear Pathology Following Lateral Wall and Peri-modiolar Cochlear Implantation.](#)

[Shaul C](#), [Bester CW](#), [Weder S](#), [Choi J](#), [Eastwood H](#), [Padmavathi KV](#), [Collins A](#), [O'Leary SJ](#).

OBJECTIVES/HYPOTHESIS: Spikes in cochlear implant impedance are associated with inner ear pathology after implantation. Here, we correlate these spikes with episodes of hearing loss and/or vertigo, with a comparison between lateral wall and peri-modiolar electrode arrays.

METHODS: Seven hundred seventy recipients of Cochlear's slim-straight, lateral wall electrode (CI422), or peri-modiolar (CI512) electrode were investigated for impedance spikes. Impedance fluctuations were defined as a median rise of ≥ 4 k Ω across all intracochlear electrodes from baseline measurements taken 2 weeks after switch-on. Medical records were analyzed from 189 of the 770 patients.

RESULTS: The slim straight, lateral wall electrode was found to spike in impedance at a significantly higher rate than the peri-modiolar array (17% vs 12%). The peri-modiolar electrode tended to spike in impedance earlier than the slim-straight electrode. Impedance spikes were found to significantly correlate with medical events (hearing loss, vertigo, or tinnitus). Overall, in the "spike" group, 42 of 75 patients (56%) demonstrated a clinical event during the impedance spike, whereas 26 of 114 patients (22%) of the "non-spike" group had a clinical event. This significant difference existed with both implant types.

CONCLUSION: These results demonstrate a small, but significant increase in impedance spikes in lateral wall electrodes, and support the relationship between spikes in cochlear implant impedances and postoperative inner-ear events, including the loss of residual hearing and vertigo. Monitoring cochlear implant impedance may be a method for early detection, and so the prevention, of these events in the future.

Genet Med. 2019 Nov;21(11):2614-2630. doi: 10.1038/s41436-019-0563-5. Epub 2019 Jun 7.

[A proposal for comprehensive newborn hearing screening to improve identification of deaf and hard-of-hearing children.](#)

[Shearer AE](#), [Shen J](#), [Amr S](#), [Morton CC](#), [Smith RJ](#); [Newborn Hearing Screening Working Group of the National Coordinating Center for the Regional Genetics Networks](#).

Early intervention for newborns who are deaf or hard-of-hearing leads to improved language, communication, and social-emotional outcomes. Universal physiologic newborn hearing screening has been widely implemented across the United States with the goal of identifying newborns who are deaf or hard-of-hearing, thereby

reducing time to diagnosis and intervention. The current physiologic newborn hearing screen is generally successful in accomplishing its goals but improvements could be made. In the past ten years, genetic testing has emerged as the most important etiologic diagnostic test for evaluation of children with deafness and congenital cytomegalovirus has been recognized as a major cause of childhood deafness that may be treatable. A comprehensive newborn hearing screen that includes physiologic, genetic, and cytomegalovirus testing would have multiple benefits, including (1) identifying newborns with deafness missed by the current physiologic screen, (2) providing etiologic information, and (3) possibly decreasing the number of children lost to follow up. We present a framework for integrating limited genetic testing and cytomegalovirus screening into the current physiologic newborn hearing screening. We identify needed areas of research and include an overview of genome sequencing, which we believe will become available over the next decade as a complement to universal physiologic newborn hearing screening.

Cold Spring Harb Perspect Med. 2019 Sep 3;9(9). pii: a033258. doi: 10.1101/cshperspect.a033258.

[The Epidemiology of Deafness.](#)

[Sheffield AM](#), [Smith RJH](#).

Hearing loss is the most common sensory deficit worldwide. It affects ~5% of the world population, impacts people of all ages, and exacts a significant personal and societal cost. This review presents epidemiological data on hearing loss. We discuss hereditary hearing loss, complex hearing loss with genetic and environmental factors, and hearing loss that is more clearly related to environment. We also discuss the disparity in hearing loss across the world, with more economically developed countries having overall lower rates of hearing loss compared with developing countries, and the opportunity to improve diagnosis, prevention, and treatment of this disorder.

Eur J Clin Microbiol Infect Dis. 2019 Dec;38(12):2365-2369. doi: 10.1007/s10096-019-03689-7. Epub 2019 Sep 3.

[The evaluation of the appropriate gentamicin use for preterm infants.](#)

[Shimizu D](#), [Ichikawa S](#), [Hoshina T](#), [Kawase M](#), [Tanaka K](#), [Araki S](#), [Kinjo T](#), [Kusuhara K](#).

Gentamicin (GM) is used for neonates as the initial treatment for neonatal bacterial infection. An association between high trough GM levels and the elevation of the serum creatinine (sCr) level and hearing loss has been reported, although there have been no reports investigating the serial changes in the sCr level in preterm neonates treated with GM. The present study evaluated the serial changes in the sCr level and the incidence of hearing loss in preterm neonates treated with GM. This study included 56 neonates born at a gestational age of 32-36 weeks. Fifteen (group 1) and 20 (group 2) neonates were treated with 2.5 mg/kg of GM every 12 h and 4 mg/kg of GM every 36 h, respectively. Group 3 included 21 neonates without GM therapy. Serum GM levels, serial changes in the sCr levels, and the incidence of hearing loss were then compared among the three groups. The serum trough GM level in group 2 was significantly lower than that in group 1 ($P < 0.001$), whereas the serum peak GM levels in these groups were almost the same. The ratio of the sCr level at birth to that at the 5th day of life in group 1 was the lowest among the 3 groups. No neonates had hearing loss. GM therapy worsened the sCr level in late preterm neonates, especially those with multiple doses per day. The appropriate use of GM is needed in order to prevent the occurrence of nephrotoxicity.

Int J Dev Neurosci. 2019 Aug;76:34-40. doi: 10.1016/j.ijdevneu.2019.05.009. Epub 2019 Jun 4.

[The left lateral occipital cortex exhibits decreased thickness in children with sensorineural hearing loss.](#)

[Shiohama T](#), [McDavid J](#), [Levman J](#), [Takahashi E](#).

Patients with sensorineural hearing loss (SNHL) tend to show language delay, executive functioning deficits, and visual cognitive impairment, even after intervention with hearing amplification and cochlear implants, which suggest altered brain structures and functions in SNHL patients. In this study, we investigated structural brain MRI in 30 children with SNHL (18 mild to moderate [M-M] SNHL and 12 moderately severe to profound [M-P] SNHL) by comparing gender- and age-matched normal controls (NC). Region-based analyses did not show statistically significant differences in volumes of the cerebrum, basal ganglia, cerebellum, and the ventricles between SNHL and NC. On surface-based analyses, the global and lobar cortical surface area, thickness, and volumes were not statistically significantly different between SNHL and NC participants. Regional surface areas, cortical thicknesses, and cortical volumes were statistically significantly smaller in M-P SNHL compared to NC in the left middle occipital cortex, and left inferior occipital cortex after a correction for multiple comparisons using random field theory ($p < 0.02$). These regions were identified as areas known to be related to high level visual cognition including the human middle temporal area, lateral occipital area, occipital face area, and V8. The observed regional decreased thickness in M-P SNHL may be associated with dysfunctions of visual cognition in SNHL detectable in a clinical setting.

J Virol Methods. 2020 Jan;275:113759. doi: 10.1016/j.jviromet.2019.113759. Epub 2019 Oct 31.

Evaluation of saliva pools method for detection of congenital human cytomegalovirus infection.

[Silva J](#), [Fernandes C](#), [Marques A](#), [Maria AT](#), [Correia C](#), [Tuna ML](#), [Chasqueira MJ](#), [Paixão P](#).

Human cytomegalovirus (HCMV) is the most frequent cause of congenital infection, leading to long-term sequelae especially sensorineural hearing loss (SNHL). Since 5-15 % of the asymptomatic newborns will develop late sequelae, the implementation of a universal screening would allow the identification of infected children and early intervention. The aim of this study was to validate the use of saliva pools of 10 and 20 samples for the detection of HCMV congenital infection. Four spiking samples (negative saliva matrix added with known concentration of AD169 strain culture supernatant) and a set of 12 saliva samples, collected from newborns with confirmed congenital infection in their first three weeks of life, were tested individually and after dilution in 10 and 20 pools by an "in-house" RT-PCR. Both pool methodologies, 10-pool and 20-pool samples, had 100 % sensitivity and specificity when compared with individual samples. This methodology could allow a cost reduction close to 85 % and 89 %, respectively for the 10-pool and 20-pool approach, when compared with testing each sample individually. This significant reduction may open the possibility to perform the newborn screening for HCMV in a large-scale.

Eur Ann Otorhinolaryngol Head Neck Dis. 2019 Oct;136(5):385-391. doi: 10.1016/j.anorl.2019.05.018. Epub 2019 Jun 17.

Guidelines (short version) of the French Society of Otorhinolaryngology (SFORL) on pediatric cochlear implant indications.

[Simon F](#), [Roman S](#), [Truy E](#), [Barone P](#), [Belmin J](#), [Blanchet C](#), [Borel S](#), [Charpiot A](#), [Coez A](#), [Deguine O](#), [Farinetti A](#), [Godey B](#), [Lazard D](#), [Marx M](#), [Mosnier I](#), [Nguyen Y](#), [Teissier N](#), [Virole B](#), [Lescanne E](#), [Loundon N](#).

OBJECTIVES: The authors present the guidelines of the French Society of Otorhinolaryngology - Head and Neck Surgery (Société française d'oto-rhino-laryngologie et de chirurgie de la face et du cou - SFORL) on the indications for cochlear implantation in children.

METHODS: A multidisciplinary work group was entrusted with a review of the scientific literature on the above topic. Guidelines were drawn up, based on the articles retrieved and the group members' individual experience. They were then read over by an editorial group independent of the work group. The guidelines were graded as A, B, C or expert opinion, by decreasing level of evidence.

RESULTS: The SFORL recommends that children with bilateral severe/profound hearing loss be offered bilateral cochlear implantation, with surgery before 12months of age. In sequential bilateral cochlear implantation in children with severe/profound hearing loss, it is recommended to reduce the interval between the two implants, preferably to less than 18months. The SFORL recommends encouraging children with unilateral cochlear implants to wear contralateral hearing aids when residual hearing is present, and recommends assessing perception with hearing-in-noise tests. It is recommended that the surgical technique should try to preserve the residual functional structures of the inner ear as much as possible.

Int J Pediatr Otorhinolaryngol. 2019 Sep 10;128:109676. doi: 10.1016/j.ijporl.2019.109676. [Epub ahead of print]

Prevalence of hearing loss among polish school-age children from rural areas - Results of hearing screening program in the sample of 67 416 children.

[Skarżyński H](#), [Gos E](#), [Świerniak W](#), [Skarżyński PH](#).

BACKGROUND: Hearing loss in children is a relevant health issue, both for its prevalence and for its physical, emotional and social consequences. Our aim was to estimate the national prevalence of hearing loss in school-age children from rural areas in Poland.

METHODS: The study was conducted in the general, pediatric, nonclinical population of school-age children from rural areas in Poland. It was a population-based, epidemiological study. The participants were 67416 children (32630 girls and 34786 boys) aged from 6 to 13 years old (M=8.65; SD=2.54). Pure-tone air-conduction hearing threshold were obtained at 0.5-8 kHz. Hearing loss was defined as a pure-tone average higher than 20 dB in one or both ears in at least one of the following pure-tone average: four-frequency pure-tone average (FFPTA), high-frequency pure-tone average (HFPTA) and low-frequency pure-tone average (LFPTA).

RESULTS: The rate of positive results of hearing screening was 16.4% and it was significantly higher in younger children than in older children. Mild hearing loss was more frequently than moderate or worse hearing loss. The children more often experienced unilateral than bilateral hearing loss.

CONCLUSIONS: This study reveals that hearing problems are common in this population, especially among younger children. It shows a strong need for systematic monitoring of hearing status among children and

increasing awareness of parents and educators of the significance of hearing loss, including unilateral and mild hearing loss. Further studies conducted among children in urban areas are needed to compare the prevalence of hearing loss in children from various environments.

Int J Pediatr Otorhinolaryngol. 2019 Aug;123:69-74. doi: 10.1016/j.ijporl.2019.04.032. Epub 2019 Apr 24.

[**The emergence and prevalence of hearing loss in children with homozygous sickle cell disease.**](#)

[Stuart A, Smith MR.](#)

OBJECTIVES: Many children with sickle cell disease (SCD) experience the effect of cochlear insult due to hemolytic anemia and vaso-occlusion that accompanies the disease. As a result, hearing sensitivity eventually decreases. The purpose of this study was to investigate the emergence of hearing loss and the prevalence of such in children with homozygous SCD.

METHODS: A large pediatric database was utilized to gather audiometric outcomes in children with homozygous SCD. Children were identified by primary diagnosis ICD codes. Demographic and audiometric data was drawn to determine presence, type, degree, and laterality of hearing loss. Hearing loss was defined as an elevated audiometric threshold >15 dB HL for pure tone air conduction audiometry or >20 dB HL minimal response level for sound-field testing.

RESULTS: One hundred and twenty-eight children were identified. The prevalence of hearing loss ranged from 28.8% to 50.8% depending on the calculation method (i.e., individual vs. ear specific prevalence and any elevated threshold vs. a three-frequency pure tone average). Conductive hearing loss and bilateral loss were most prevalent. The degree of hearing loss ranged from slight to profound. The mean age of identification of sensorineural hearing loss was 9.6 years.

CONCLUSION: The prevalence of hearing loss in children with homozygous SCD is higher than in normal children. Regular hearing assessments of children with SCD is warranted and should be advocated in early infancy and as part of their ongoing care.

Eur Arch Otorhinolaryngol. 2019 Sep;276(9):2433-2439. doi: 10.1007/s00405-019-05498-2. Epub 2019 Jun 7.

[**Clinical benefit of wideband-tympanometry: a pediatric audiology clinical study.**](#)

[Stuppert L, Nospes S, Bohnert A, Läßig AK, Limberger A, Rader T.](#)

PURPOSE: Wideband-tympanometry (WBT) could give more informative data about the tympanic condition than the conventional tympanometry. In the actual literature, the clinical profit of wideband-tympanometry in pediatric audiological settings is not well evaluated. The aim of this study was to analyze the additional clinical benefit.

METHODS: 150 children (281 ears) with normal hearing, at the age from 11 days up to 14;10 years, checked with pure tone audiometry or auditory brainstem responses (ABR) participated in this retrospective study. We divided in four age ranges (≤ 6 month; > 6 month ≤ 3 years; > 3 years ≤ 11 years; > 11 years). All children were evaluated with ENT examination including ear microscopy, conventional 226-Hz or 1000-Hz tympanometry and WBT. Ear canal volumes were determined.

RESULTS: Compared with literature data, our patients aged ≤ 3 years showed smaller mean ear canal volumes (≤ 4 ml). We found a good statistical correlation between the WBT-results and 1000-Hz tympanometry but a rare correlation between WBT-results and ear microscopic findings. In the patients with pathologic ear microscopic results in all groups of age, a significant reduction of WBT-absorbance in 1000 Hz and 2000 Hz was found.

CONCLUSIONS: This study confirms that WBT collects additive data to detect the correct middle ear status. In pediatric audiology, WBT is an additional useful method to value middle ear problems and to analyze the character of infantile hearing loss. Standard guidelines for the interpretation of the pediatric population are needed. Hence, it will be necessary to determine these findings in a larger number of infantile ears.

Aust N Z J Public Health. 2019 Oct 30. doi: 10.1111/1753-6405.12948. [Epub ahead of print]

[**The impact of hearing impairment on Aboriginal children's school attendance in remote Northern Territory: a data linkage study.**](#)

[Su JY, He VY, Guthridge S, Howard D, Leach A, Silburn S.](#)

OBJECTIVE: To investigate the association between hearing impairment (HI) and Year 1 school attendance in Aboriginal children in the Northern Territory (NT) of Australia.

METHODS: Observational cohort study ($n=3,744$) by analysing linked individual-level information for Aboriginal children from the NT Government school attendance records, NT Perinatal Register and Remote Hearing Assessment dataset, and community level data for relative remoteness, socioeconomic disadvantage and housing crowdedness.

RESULTS: Children with unilateral hearing loss, mild HI and moderate or worse HI had significantly lower Year 1

attendance than those with normal hearing, attending 5.6 (95%CI, -9.10 ~-2.10), 4.0 (95%CI, -7.17 ~-0.90) and 6.1 (95%CI, -10.71 ~-1.49) days fewer, respectively. Other variables that yielded significant association were: male gender, having attended preschool less than 20% of available days, speaking English as second language, twin birth and average household size >5.

CONCLUSIONS: Aboriginal children with any level of HI are likely to have lower school attendance rates in Year 1 than their peers with normal hearing. Implications for public health: In this population, where the prevalence of otitis media and accompanying HI remains extremely high, the early detection and management of hearing loss on entry into primary school should be included in the measures to improve school attendance.

Eye (Lond). 2019 Jul 18. doi: 10.1038/s41433-019-0536-6. [Epub ahead of print]

Retinal findings in pediatric patients with Usher syndrome Type 1 due to mutations in MYO7A gene.

[Subirà O](#), [Català-Mora J](#), [Díaz-Cascajosa J](#), [Padrón-Pérez N](#), [Claveria MA](#), [Coll-Alsina N](#), [Bonnet C](#), [Petit C](#), [Caminal JM](#), [Prat J](#).

PURPOSE: To describe retinal alterations detected by swept-source optical coherence tomography (SS-OCT) in paediatric patients with Usher syndrome type 1 (USH1) and to compare these findings to previously published reports.

METHODS: Thirty-two eyes from 16 patients (11 males and 5 females) with a genetic diagnosis of USH1 because of MYO7A mutations underwent SS-OCT. Patients ranged in age from 4 to 17 years (mean, 11,13±4,29). The subfoveal and macular area were analysed with SS-OCT at 1050 nm using 12 radial scans of 12.0 mm. Structural abnormalities were evaluated and correlated with best-corrected visual acuity (BCVA).

RESULTS: The most common qualitative retinal abnormality was external layer damage in macular area. Specific alterations included external limiting membrane loss/disruption (27 eyes; 84.4%), disruption of the Myoid zone (27 eyes; 84.4%); Ellipsoid zone disruption (28 eyes; 87.5%), and loss of the outer segments (29 eyes; 90.6%). The damage of the retinal pigment epithelium was divided according to the loss of the different layers: phagosome zone (30 eyes; 93.8%), melanosome zone (29 eyes; 90.6%) and mitochondria zone (0 eyes; 0%). The presence of cystoid macular oedema (CMO) was significantly correlated with alterations in photoreceptors. Disruption or absence of the myoid and ellipsoid zones of the photoreceptors were the only variables independently associated with decreased BCVA.

CONCLUSIONS: The findings of this study suggest that the physiopathologic basis of early-stage Usher syndrome (USH) may be changes in the outer retinal layer, particularly the photoreceptors, which in turn may cause alterations-such as CMO-in the inner retinal layers. Accordingly, monitoring the condition of photoreceptors during follow-up may be advisable for the early detection of pathologic changes.

Cochlear Implants Int. 2019 Oct 7:1-9. doi: 10.1080/14670100.2019.1670486. [Epub ahead of print]

Self-reported hearing quality of life measures in pediatric cochlear implant recipients with bilateral input.

[Suneel D](#), [Davidson LS](#), [Lieu J](#).

OBJECTIVE: Self-reported hearing quality of life (QoL) for pediatric cochlear implant (CI) recipients was examined, asking whether 1) children with CIs have similar QoL as those with less severe hearing loss (HL); 2) children with different bilateral CI (BCI) device configurations report different QoL; and 3) do audiological, demographic and spoken language factors affect hearing QoL?

DESIGN: One hundred four children (ages 7-11 years) using bimodal devices or BCIs participated. The Hearing Environments and Reflection of Quality of Life (HEAR-QL) questionnaire, receptive language and speech perception tests were administered. HEAR-QL scores of CI recipients were compared to scores of age-mates with normal hearing and mild to profound HL.

RESULTS: HEAR-QL scores for CI participants were similar to those of children with less severe HL and did not differ with device configuration. Emotion identification and word recognition in noise correlated significantly with HEAR-QL scores.

DISCUSSION: CI recipients reported that HL hinders social participation. Better understanding of speech in noise and emotional content was associated with fewer hearing-related difficulties on the HEAR-QL.

CONCLUSIONS: Noisy situations encountered in educational settings should be addressed for children with HL. The link between perception of emotion and hearing-related QoL for CI recipients should be further examined.

Bacterial profile and antibacterial susceptibility of otitis media among pediatric patients in Hawassa, Southern Ethiopia: cross-sectional study.

Tadesse B, Shimelis T, Worku M.

BACKGROUND: Otitis Media (OM) is the most common disease of childhood. Twenty thousand people die each year from otitis media. It is an important cause of preventable hearing loss, affects children's intellectual performance and language development. There are very small numbers of studies done in Ethiopia concerning this topic. This study aimed to identify bacterial pathogens related to ear infection and to assess antibacterial susceptibility of isolated organisms.

METHOD: A cross-sectional study was conducted on 152 children from April 2018 to July 2018 at selected health facilities in Hawassa city, SNNPR, Ethiopia. All pediatric patients having ear discharge were included. Convenient sampling technique was used to collect clinical and demographic data using standard questionnaires after child care-takers signed the consent. Ear discharge specimens were collected using a sterile swab, and transported using Amies transport media to Hawassa University Comprehensive Specialized Hospital laboratory. Bacterial isolates were characterized based on colony appearance, Gram reaction, culture characteristics, and biochemical tests after inoculating on appropriate culture media. Antibacterial susceptibility testing was performed using the disc diffusion method according to the criteria of the Clinical and Laboratory Standards Institute (CLSI).

RESULTS: Among 152 children included, 115(75.6%) of them demonstrated pathogenic bacterial growth. *Staphylococcus aureus* 41(27%) was the most frequently isolated pathogen, followed by *Proteus mirabilis* 19 (12.5%). Of the total isolates, 11.2 and 7.3% were resistant to gentamicin and ciprofloxacin respectively. Over three-fourth (85.2%) of the isolates were resistant to ampicillin. More than two-third of the isolates were resistant to both penicillin (71.4%) and trimethoprim-sulphamethoxazole (72.0%).

CONCLUSIONS: *S. aureus* is the most commonly isolated bacterial pathogen from ear discharge among children. Even though gentamicin is a parenteral drug and ciprofloxacin is rarely used in children due to concerns of bone/joint effects, these two drugs were highly effective antibiotics and thus should be considered in treating children with otitis media since most organisms were resistance or poor response to first line drugs. High level of antibiotic resistance was observed so antimicrobial susceptibility test is needed before prescribing drugs for treatment of OM.

Pediatric cochlear implantation: A quarter century in review.

Teagle HFB, Park LR, Brown KD, Zdanski C, Pillsbury HC.

OBJECTIVE: To review the growth of a pediatric cochlear implant (CI) program at one large tertiary care medical center over a 25-year period in order to (1) describe the population of pediatric cochlear implant recipients, (2) document word recognition outcomes, and (3) describe changes in candidacy criteria over time.

DESIGN: A retrospective review of population demographics and trends included etiology of hearing loss, device use and type, expansion of inclusion criteria, and word recognition outcomes.

RESULTS: Ninety-one percent of the children studied were from North Carolina and reflect the ethnic distribution of the state. The population is heterogeneous for etiology and the presence of syndromes and/or co-morbidities. A trend of lower age of implant and greater residual hearing was documented overtime. As a single metric, monosyllable word recognition for the children assessed is good with the mean CNC test word score of 76.13% (range 0-100, S.D=19.94).

CONCLUSIONS: Pediatric cochlear implant candidacy criteria have evolved despite no change in FDA-approved regulations since 2000. There is great diversity among recipients but word recognition outcomes are generally good in this population and have improved over time. Professionals who may refer children for cochlear implantation should be aware of current clinical practices and general outcomes.

Hearing Thresholds in Young Children With Otitis Media With Effusion With and Without Cleft Palate.

Tengroth B, Lohmander A, Hederstierna C.

OBJECTIVE: To investigate hearing thresholds in children born with cleft palate and in children with otitis media with effusion but no cleft palate.

DESIGN: Prospective longitudinal group comparison study.

SETTING: University hospital.

PARTICIPANTS: Sixteen children born with nonsyndromic cleft palate with or without cleft lip (CP±L) and 15 age-

matched children with otitis media with effusion (OME) but without cleft.

MAIN OUTCOME MEASURES: Hearing was tested at repeated occasions beginning with neonatal auditory brainstem response (ABR) at 1-4 months of age, and age-appropriate hearing tests from 9 to 36 months of age.

RESULTS: The median ABR thresholds in both groups were elevated but did not differ significantly. At 12 months of age, the median 4 frequency averages at 500-1000-2000-4000 Hz (4FA) were indicative of mild hearing loss but significantly better in the CP±L-group than in the group without cleft ($P < .01$). There were no significant group-wise differences regarding the median 4FA at 24 and 36 months of age, and at 36 months, the median 4FA were normal in both groups. Both groups exhibited a significant improvement over time from the neonatal ABR thresholds to the 4FA at 36 months (CP±L-group $P < .05$; without CP±L-group $P < .01$).

CONCLUSION: The hearing loss in children with CP±L was not more severe than among children with OME but without cleft palate; rather, at 12 months of age, the thresholds were significantly better in the CP±L-group than those in the group without cleft. The air conduction thresholds improved with age in both groups.

Acta Paediatr. 2019 Nov;108(11):1972-1977. doi: 10.1111/apa.14837. Epub 2019 Jun 2.

Prevalence and risk factors for hearing loss in high-risk neonates in Germany.

Thangavelu K, Martakis K, Fabian S, Venkateswaran M, Roth B, Beutner D, Lang-Roth R.

AIM: Hearing loss in infants is often diagnosed late, despite universal screening programmes. Risk factors of hearing impairment in high-risk neonates, identified from population-based studies, can inform policy around targeted screening. Our aim was to determine the prevalence and the risk factors of hearing loss in a high-risk neonatal population.

METHODS: This was a retrospective cohort study of neonates hospitalised at the University Hospital Cologne, Germany from January 2009 to December 2014 and were part of the newborn hearing screening programme. Multivariable regression analyses using the lasso approach was performed.

RESULTS: Data were available for 4512 (43% female) neonates with a mean gestational age at birth of 35.5 weeks. The prevalence of hearing loss was 1.6%, and 42 (0.9%) neonates had permanent hearing loss. Craniofacial anomalies, hyperbilirubinaemia requiring exchange transfusion, oxygen supplementation after 36 weeks of gestation and hydrops fetalis showed associations with permanent hearing loss.

CONCLUSION: Our findings of risk factors for hearing loss were consistent with other studies. However, some commonly demonstrated risk factors such as perinatal infections, meningitis, sepsis and ototoxic drugs did not show significant associations in our cohort. Targeted screening based on risk factors may help early identification of hearing loss in neonates.

Otol Neurotol. 2019 Nov 15. doi: 10.1097/MAO.0000000000002457. [Epub ahead of print]

Electrode Array Type and Its Impact on Impedance Fluctuations and Loss of Residual Hearing in Cochlear Implantation.

Thompson NJ, Dillon MT, Buss E, Park LR, Pillsbury HC 3rd, O'Connell BP, Brown KD.

HYPOTHESIS/OBJECTIVE: Determine variables associated with electrode impedance fluctuations and loss of residual hearing in cochlear implant (CI) recipients.

BACKGROUND: CI recipients with postoperative hearing preservation demonstrate superior speech perception with an electric-acoustic stimulation (EAS) device as compared with a CI-alone device. Maintaining superior speech perception over time relies on long-term hearing preservation; therefore, understanding variables that may contribute to loss of residual hearing is needed. Recent reports suggest a relationship between changes in electrode impedance and loss of residual hearing. The variables influencing this relationship have yet to be determined.

METHODS: Review of pediatric and adult CI cases from 2013 to 2016 who presented with preoperative residual hearing. Regression analysis was performed to evaluate effects of array type (lateral wall vs. perimodiolar), manufacturer, age at implantation, and preoperative hearing on impedance. The correlation between peak impedance change and change in low-frequency hearing was determined.

RESULTS: One hundred forty-six CI recipients presented with preoperative residual hearing. A multivariate regression analysis demonstrated a statistically significant association between preoperative hearing thresholds ($p=0.017$), device manufacturer ($p=0.011$), and array type ($p=0.038$) on postoperative impedance changes. Hearing preservation rates and change in impedance differed by electrode array type. The association between peak impedance changes and loss of residual hearing differed between manufacturers ($R=0.208$, $p=0.029$ vs. $R=0.016$, $p=0.609$).

CONCLUSION: Impedance fluctuation appears to be a marker for loss of residual hearing for specific electrode array types and manufacturers. Specific arrays may affect the cochlear microenvironment differently, with different effects on postoperative hearing preservation.

Laryngoscope. 2019 Nov;129(11):E407-E411. doi: 10.1002/lary.28150. Epub 2019 Jul 3.

Idiopathic Intracranial Hypertension Presenting as Auditory Neuropathy Hearing Disorder in a Child.

Tobia A, Khayr R, Shpak T, Peleg C, Genizi J, Khnifes R, Borissovsky N, Luntz M.

Otologic manifestations are known to occur in patients with idiopathic intracranial hypertension (IIH), but the occurrence of sensorineural hearing loss, especially in pediatric populations, has been addressed in only a few reports. Here, we describe a pediatric patient who presented with IIH and severe bilateral hearing loss. The patient's hearing loss was diagnosed as a form of auditory neuropathy (AN) and resolved after prompt treatment of the increased intracranial pressure. This case points to a possible association between IIH and AN and suggests that IIH may potentially be a reversible cause of AN spectrum disorder. Laryngoscope, 129:E407-E411, 2019.

Otolaryngol Head Neck Surg. 2019 Jul;161(1):150-156. doi: 10.1177/0194599819836679. Epub 2019 Mar 12.

Pediatric Stapes Surgery: Hearing and Surgical Outcomes in Endoscopic vs Microscopic Approaches.

Tolisano AM, Fontenot MR, Nassiri AM, Hunter JB, Kutz JW Jr, Rivas A, Isaacson B.

OBJECTIVE: To compare endoscopic and microscopic pediatric stapes surgery.

STUDY DESIGN: Case series with chart review.

SETTING: Two academic otology practices.

SUBJECTS AND METHODS: Surgical and hearing outcomes were compared for consecutive children (<18 years) undergoing microscopic and endoscopic stapes surgery. The main outcome measure was closure of the air-bone gap (ABG) to ≤ 20 dB.

RESULTS: Twenty-two endoscopic surgeries (17 stapedectomies, 4 stapedotomies, and 1 stapes mobilization) and 52 microscopic surgeries (30 stapedectomies, 19 stapedotomies, and 3 stapes mobilizations) were performed. Patient demographics, history of ipsilateral middle ear surgery, and revision stapes surgery status were similar. The most common diagnosis for the endoscopic group and microscopic group were congenital stapes footplate fixation (45.5%) and juvenile otosclerosis (46.2%), respectively. Preoperative ABGs in the endoscopic (37.7 dB) and microscopic (32.8 dB) groups ($P = .170$) were similar. There were no major complications, including facial nerve injury or anacusis, in the endoscopic group. Postoperative sensorineural hearing loss (>15 dB) did not occur in any patients in the endoscopic group but was present in 2 patients in the microscopic group ($P = .546$). Improvement in pure-tone average (25.9 dB vs 18.5 dB, $P = .382$) and ABG (21.7 dB vs 14.7 dB, $P = .181$) was similar, and postoperatively, the median ABG was 11.3 dB and 15.0 dB for endoscopic and microscopic cases ($P = .703$), respectively. ABG closure to ≤ 20 dB (72.7% vs 65.2%, $P = .591$) was also similar.

CONCLUSION: Pediatric endoscopic stapes surgery is safe and hearing outcomes are similar to the microscopic approach when performed by experienced endoscopic ear surgeons.

BMC Pregnancy Childbirth. 2019 Jun 20;19(1):205. doi: 10.1186/s12884-019-2360-1.

Serological screening of immunoglobulin M and immunoglobulin G during pregnancy for predicting congenital cytomegalovirus infection.

Torii Y, Yoshida S, Yanase Y, Mitsui T, Horiba K, Okumura T, Takeuchi S, Suzuki T, Kawada JI, Kotani T, Yamashita M, Ito Y.

BACKGROUND: Cytomegalovirus (CMV) is one of the most frequent pathogens for congenital infections. Most cases of congenital CMV infection (cCMV) are asymptomatic at birth, but sensorineural hearing loss (SNHL) or neurodevelopmental delay can appear later in childhood. This prospective study examined the practicability of serological screening for anti-CMV immunoglobulin (Ig) G and anti-CMV IgM in pregnant women.

METHODS: A total of 11,753 pregnant women were examined for CMV IgG and CMV IgM during the first or second trimester. When IgM was positive, IgG was reevaluated more than two weeks later. When IgG was negative, IgG was reevaluated in the second or third trimester. All neonates from mothers with positive/borderline IgM or IgG seroconversion underwent polymerase chain reaction assay for CMV using urine samples to diagnose cCMV. Levels of IgG and IgM were compared between mothers with and without cCMV. Receiver operating characteristic (ROC) curves for IgM titers were analyzed.

RESULTS: Eight of 500 neonates (1.6%) born from mothers with positive IgG and positive IgM, and 3 of 13 neonates (23.1%) born from mothers with IgG seroconversion were diagnosed with cCMV. Neither IgM titers nor IgG titers differed significantly between cCMV and non-cCMV groups. The area under the ROC curve was 0.716 and the optimal cut-off for IgM was 7.28 index (sensitivity = 0.625, specificity = 0.965, positive predictive value = 0.238, negative predictive value = 0.993). Titers of IgG were not frequently elevated in pregnant

women with positive IgM during the observation period, including in those with cCMV. All 11 cCMV cases were asymptomatic at birth and none had shown SNHL or developmental delay as of the last regular visit (mean age, 40 months).

CONCLUSIONS: Seroconversion of CMV IgG and high-titer IgM during early pregnancy are predictors of cCMV. High IgM titer (>7.28 index) is a predictor despite relatively low sensitivity. Levels of IgG had already plateaued at first evaluation in mothers with cCMV. Maternal screening offered insufficient positive predictive value for diagnosing cCMV, but allowed identifying asymptomatic cCMV cases in an early stage.

Cancer Chemother Pharmacol. 2019 Dec;84(6):1333-1338. doi: 10.1007/s00280-019-03968-2. Epub 2019 Oct 4.

Cisplatin ototoxicity in children: risk factors and its relationship with polymorphisms of DNA repair genes ERCC1, ERCC2, and XRCC1.

Turan C, Kantar M, Aktan Ç, Kosova B, Orman M, Bilgen C, Kirazlı T.

PURPOSE: We aimed to investigate the cisplatin-related hearing toxicity and its possible relationship with polymorphic variants in DNA repair genes, ERCC1, ERCC2, and XRCC1.

METHODS: Fifty patients treated with cisplatin in the past were included in the study. There were 29 females and 21 males; mean age 13.4 ± 6.0 years). The polymorphism in DNA repair genes was studied using primer and probes in Light Cycler device after DNA isolation was carried out with PCR technique. The polymorphisms and clinical risk factors were evaluated using Chi square test and logistic regression modelling.

RESULTS: The patients had hearing loss in 44%. For ERCC1 gene, the patients with hearing loss had 50% of GG (wild type), 40.9% of AG and 9.1% of AA genotypes, while the patients without hearing loss had 28.6% of GG, 53.5% of AG, and 17.9% of AA genotypes. For ERCC2 gene, the patients with hearing loss had 18.2% of GG (wild type), 40.9% of TG, and 40.9% of TT genotypes, while the patients without hearing loss had 10.7% of GG 39.3% of TG, and 50% of TT genotypes. For XRCC1 gene, the patients with hearing loss had 18.2% of CC (wild type), 59.1% of CT, and 22.7% of TT genotypes, while the patients without hearing loss had 35.7% of CC, 50% of CT, and 14.3% of TT genotypes. There was no statistically significant association among the groups ($p=0.24$).

CONCLUSION: We did not find a relationship between DNA repair gene polymorphisms and hearing toxicity of cisplatin.

Int J Pediatr Otorhinolaryngol. 2019 Nov;126:109614. doi: 10.1016/j.ijporl.2019.109614. Epub 2019 Jul 29.

First experiences with a new adhesive bone conduction hearing device in children.

Urík M, Hošnová D, Šlapák I, Jančíková J, Odstrčilík J, Jarkovský J, Baumgartner WD.

OBJECTIVES: To evaluate the hearing benefit, advantages, and disadvantages in a series of children using a new, nonimplantable, pressure-free, adhesive bone conduction hearing aid.

METHODS: Seventeen children were included in the study. 5 children suffered from bilateral conductive hearing loss (CHL), 6 children with unilateral CHL and 6 children with unilateral sensorineural hearing loss. An audiological tests were provide. Additionally, sound quality (SSQ10) and quality of life (AQoL-6D) were assessed using questionnaires.

RESULTS: The average value of speech audiometry with bubble noise in children with SNHL is $21.33 (\pm 5.72)$ dB HL with the device and $27.67 (\pm 4.59)$ dB HL without the device, which is a statistically significant gain ($p=0.027$). The analysis showed the average value of hearing threshold in sound field in the group of children with CHL supported $20.23 (\pm 16.84)$ dB HL and not supported $33.52 (\pm 27.27)$ by the hearing aid for bone conduction, which is a statistically significant gain ($p=0.008$). The average value of speech audiometry is $23.45 (\pm 14.45)$ dB HL with the device and $37.27 (\pm 26.65)$ dB HL without the device, which is a statistically significant gain ($p=0.012$). The average value of speech audiometry with bubble noise is $30.55 (\pm 10.03)$ dB HL with the device and $45.45 (\pm 18.41)$ dB HL without the device, which is a statistically significant gain ($p=0.008$). No patient referred pain or irritation.

CONCLUSION: This new device for bone conduction show a hearing benefit for a paediatric patient, without any concomitant aesthetic and other complications.

Cochrane Database Syst Rev. 2019 May 7;5:CD009219. doi: 10.1002/14651858.CD009219.pub5.

Medical interventions for the prevention of 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, oxaliplatin or a combination of these, is used to treat a variety of paediatric malignancies. One of the most significant adverse effects is the occurrence of hearing loss or ototoxicity. In an effort to prevent this ototoxicity, different otoprotective medical interventions have been studied. This review is the third update of a previously published Cochrane Review.

OBJECTIVES: To assess the efficacy of medical interventions to prevent hearing loss and to determine possible effects of these interventions on antitumour efficacy, toxicities other than hearing loss and quality of life in children with cancer treated with platinum-based therapy as compared to placebo, no additional treatment or another protective medical intervention.

SEARCH METHODS: We searched the Cochrane Central Register of Controlled Trials, MEDLINE (PubMed) and Embase (Ovid) to 8 January 2019. We handsearched reference lists of relevant articles and assessed the conference proceedings of the International Society for Paediatric Oncology (2006 up to and including 2018), the American Society of Pediatric Hematology/Oncology (2007 up to and including 2018) and the International Conference on Long-Term Complications of Treatment of Children and Adolescents for Cancer (2010 up to and including 2015). We scanned ClinicalTrials.gov and the World Health Organization International Clinical Trials Registry Platform (WHO ICTRP; apps.who.int/trialsearch) for ongoing trials (on 2 January 2019).

SELECTION CRITERIA: Randomized controlled trials (RCTs) or controlled clinical trials (CCTs) evaluating platinum-based therapy with an otoprotective medical intervention versus platinum-based therapy with placebo, no additional treatment or another protective medical intervention in children with cancer.

DATA COLLECTION AND ANALYSIS: Two review authors independently performed the study selection, data extraction, risk of bias assessment and GRADE assessment of included studies, including adverse effects. We performed analyses according to the Cochrane Handbook for Systematic Reviews of Interventions.

MAIN RESULTS: We identified two RCTs and one CCT (total number of participants 149) evaluating the use of amifostine versus no additional treatment in the original version of the review; the updates identified no additional studies. Two studies included children with osteosarcoma, and the other study included children with hepatoblastoma. Children received cisplatin only or a combination of cisplatin and carboplatin, either intra-arterially or intravenously. Pooling of results of the included studies was not possible. From individual studies the effect of amifostine on symptomatic ototoxicity only (i.e. National Cancer Institute Common Toxicity Criteria version 2 (NCICTCv2) or modified Brock grade 2 or higher) and combined asymptomatic and symptomatic ototoxicity (i.e. NCICTCv2 or modified Brock grade 1 or higher) were uncertain (low-certainty evidence). Only one study including children with osteosarcoma treated with intra-arterial cisplatin provided information on tumour response, defined as the number of participants with a good or partial remission. The available-data analysis (data were missing for one participant), best-case scenario analysis and worst-case scenario analysis showed a difference in favour of amifostine, although the certainty of evidence for this effect was low. There was no information on survival for any of the included studies. Only one study, including children with osteosarcoma treated with intra-arterial cisplatin, provided data on the number of participants with adverse effects other than ototoxicity grade 3 or higher (on NCICTCv2 scale). There was low-certainty evidence that grade 3 or 4 vomiting was higher with amifostine (risk ratio (RR) 9.04, 95% confidence interval (CI) 1.99 to 41.12). The effects on cardiotoxicity and renal toxicity grade 3 or 4 were uncertain (low-certainty evidence). None of the studies evaluated quality of [life](#). In the recent update, we also identified one RCT including 109 children with localized hepatoblastoma evaluating the use of sodium thiosulfate versus no additional treatment. Children received intravenous cisplatin only (one child also received carboplatin). There was moderate-certainty evidence that both symptomatic ototoxicity only (i.e. Brock criteria grade 2 or higher) and combined asymptomatic and symptomatic ototoxicity (i.e. Brock criteria grade 1 or higher) was lower with sodium thiosulfate (combined asymptomatic and symptomatic ototoxicity: RR 0.52, 95% CI 0.33 to 0.81; symptomatic ototoxicity only: RR 0.39, 95% CI 0.19 to 0.83). The effect of sodium thiosulfate on tumour response (defined as number of participants with a complete or partial response at the end of treatment), overall survival (calculated from time of randomization to death or last follow-up), event-free survival (calculated from time of randomization until disease progression, disease relapse, second primary cancer, death, or last follow-up, whichever came first) and adverse effects other than hearing loss and tinnitus grade 3 or higher (according to National Cancer Institute Common Toxicity Criteria Adverse Effects version 3 (NCICTCAEv3) criteria) was uncertain (low-certainty evidence for all these outcomes). Quality of life was not assessed. We found no eligible studies for possible otoprotective medical interventions other than amifostine and sodium thiosulfate and for other types of malignancies.

AUTHORS' CONCLUSIONS: At the moment there is no evidence from individual studies in children with osteosarcoma or hepatoblastoma treated with different platinum analogues and dosage schedules that underscores the use of amifostine as an otoprotective intervention as compared to no additional treatment. Since pooling of results was not possible and the evidence was of low certainty, no definitive conclusions can be made. Since we found only one RCT evaluating the use of sodium thiosulfate in children with localized hepatoblastoma treated with cisplatin, no definitive conclusions on benefits and harms can be drawn. It should be noted that 'no evidence of effect', as identified in this review, is not the same as 'evidence of no effect'. We identified no eligible studies for other possible otoprotective medical interventions and other types of malignancies, so no conclusions can be made about their efficacy in preventing ototoxicity in children treated with platinum-based therapy. More high-quality research is needed.

[**The etiological evaluation of sensorineural hearing loss in children.**](#)

[van Beeck Calkoen EA](#), [Engel MSD](#), [van de Kamp JM](#), [Yntema HG](#), [Goverts ST](#), [Mulder MF](#), [Merkus P](#), [Hensen EF](#).

This study aims to evaluate the etiology of pediatric sensorineural hearing loss (SNHL). A total of 423 children with SNHL were evaluated, with the focus on the determination of causative genetic and acquired etiologies of uni- and bilateral SNHL in relation to age at diagnosis and severity of the hearing loss. We found that a stepwise diagnostic approach comprising of imaging, genetic, and/or pediatric evaluation identified a cause for SNHL in 67% of the children. The most common causative finding in children with bilateral SNHL was causative gene variants (26%), and in children with unilateral SNHL, a structural anomaly of the temporal bone (27%). The probability of finding an etiologic diagnosis is significantly higher in children under the age of 1 year and children with profound SNHL. **Conclusions:** With our stepwise diagnostic approach, we found a diagnostic yield of 67%. Bilateral SNHL often has a genetic cause, whereas in unilateral SNHL structural abnormalities of the labyrinth are the dominant etiologic factor. The diagnostic yield is associated with the age at detection and severity of hearing loss: the highest proportion of causative abnormalities is found in children with a young age at detection or a profound hearing loss. **What is Known:** • Congenital sensorineural hearing loss is one of the most common congenital disorders • Determination of the cause is important for adequate management and prognosis and may include radiology, serology, and DNA analysis **What is New:** • Using a stepwise diagnostic approach, causative abnormalities are found in 67% both in uni- and bilateral SNHL, with the highest diagnostic yield in very young children and those suffering from profound hearing loss • Bilateral SNHL often has a genetic cause, whereas in unilateral SNHL structural abnormalities of the labyrinth are the dominant etiologic factor.

[**Quality of life of children with hearing loss in special and mainstream education: A longitudinal study.**](#)

[van der Straaten TFK](#), [Rieffe C](#), [Soede W](#), [Netten AP](#), [Dirks E](#), [Oudesluys-Murphy AM](#), [Dekker FW](#), [Böhringer S](#), [Frijns JHM](#); [DECIBEL Collaborative study group](#).

OBJECTIVES: To compare the quality of life of children with hearing loss (HL) and children with normal hearing (NH) and to examine how the QoL of children with HL changes over time, considering language skills, type of hearing device, degree of HL, and type of education.

METHODS AND MATERIALS: This longitudinal study included 62 children with HL and their parents. Developmental outcome data were collected at two time points, when the mean ages of the children were 4 and 11 years. The Pediatric Quality of Life (PedsQL™) questionnaire, which includes assessments of Physical, Emotional, Social, and School functioning, was completed by parents at both time points and by the children with HL at the second time point. Receptive and expressive language skills at 4 years were assessed by the Reynell Developmental Language Scale. Results were compared with a Dutch normative sample.

RESULTS: The QoL of children with HL was similar to that of children with NH at both time points on two of the four QoL scales, Emotional and Physical functioning. On the other two scales, Social and School functioning, children with HL who attended special education and children who switched to mainstream education showed lower scores than children with HL who were consistently in mainstream education and lower scores than children with NH. The School QoL of children with HL decreased over time, as did the School QoL of children with NH. Social QoL of children with cochlear implants decreased over time, but this was not the case in children with hearing aids. Language skills and the degree of HL did not clinically improve the QoL over time of preschool children with HL.

CONCLUSIONS: The QoL of children with HL in mainstream education and the Physical and Emotional QoL of all children with HL were satisfactory. It is essential to develop specific guidance regarding school activities for children with HL in special education and for children with HL who switch to mainstream education in order to increase their social QoL.

[**Children with unilateral cochlear nerve canal stenosis have bilateral cochleovestibular anomalies.**](#)

[Vilchez-Madrigal LD](#), [Blaser SI](#), [Wolter NE](#), [James AL](#), [Papsin BC](#), [Gordon KA](#), [Cushing SL](#), [Propst EJ](#).

OBJECTIVES/HYPOTHESIS: To investigate the cochleovestibular apparatus bilaterally in children with isolated unilateral bony cochlear nerve canal (bCNC) stenosis.

STUDY DESIGN: Retrospective review.

METHODS: Imaging studies of children with unilateral bCNC stenosis (<1.0 mm) on computed tomography imaging (N=36) were compared with controls imaged due to trauma without temporal bone injury (N=32). Twenty-six measurements were obtained in each ear, assessing the bony internal auditory canal (IAC), cochlea, and vestibular end-organs, and were analyzed using one-way analysis of variance for intersubject comparisons

and paired t tests for intrasubject comparisons with a Bonferroni adjustment for multiple comparisons ($P = .0006$).

RESULTS: Patients with bCNC stenosis had a smaller IAC ($P < .000$) and cochlea ($P < .000$) on the stenotic side as compared with controls. Although the vestibular end-organ was also smaller in bCNC ears, this difference was not significant. The contralateral ear also had a smaller bCNC ($P < .000$) and cochlea ($P < .000$) as compared with controls, although to a lesser degree than the stenotic side.

CONCLUSIONS: Children with unilateral bCNC stenosis have abnormal biometry of both the cochlea and the vestibular end-organ in the affected and the normal contralateral ear as compared with controls.

Syst Rev. 2019 Jul 17;8(1):172. doi: 10.1186/s13643-019-1073-x.

[Risk factors for hearing loss in children: a systematic literature review and meta-analysis protocol.](#)

[Vos B](#), [Noll D](#), [Pigeon M](#), [Bagatto M](#), [Fitzpatrick EM](#).

BACKGROUND: Hearing loss in newborns and children is a public health concern, due to high prevalence and negative effects on their development. Early detection and intervention of childhood hearing loss may mitigate these negative effects. Population-based newborn hearing screening programs have been established worldwide to identify children at risk for congenital hearing loss and to follow children at risk for late onset or progressive hearing loss. This article presents the protocol for a systematic review that aims to review the risk factors associated with permanent hearing loss in children, including congenital, early, or late onset. Risk factors associated with progressive hearing loss will be investigated as a secondary aim.

METHODS: Scientific literature from the following databases will be investigated: MEDLINE, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R), Embase, and CINAHL. The primary outcome is a permanent bilateral or unilateral hearing loss with congenital onset or onset during childhood (birth to 18 years). The secondary outcome is progressive hearing loss. Studies must report data on risk factors associated with permanent hearing loss; risk factors may be present at birth or later and result in immediate or delayed hearing loss. Randomized controlled trials, quasi-experimental studies, nonrandomized comparative and non-comparative studies, and case series will be included. The risk of bias will be assessed using the Qualitative Assessment Tool for Quantitative Studies (McMaster University). If aggregation of data is possible for a subsection of studies, we will pool data using meta-analysis techniques. If aggregation of data is not possible, a qualitative synthesis will be presented. We will assess the quality and strength of the overall body of evidence using the Grading of Recommendations Assessment, Development and Evaluation (GRADE). The systematic review follows the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) recommendations.

DISCUSSION: The resulting information will inform the update of a provincial audiological surveillance protocol for the Ontario Infant Hearing Program and will be applicable to early hearing detection and intervention (EHDI) programs worldwide.

SYSTEMATIC REVIEW REGISTRATION: We have registered the protocol in the International Prospective Register of Systematic Reviews (PROSPERO), registration number CRD42018104121.

Front Psychol. 2019 Oct 25;10:2421. doi: 10.3389/fpsyg.2019.02421. eCollection 2019.

[Longitudinal Speech Recognition in Noise in Children: Effects of Hearing Status and Vocabulary.](#)

[Walker EA](#), [Sapp C](#), [Oleson JJ](#), [McCreery RW](#).

OBJECTIVES: The aims of the current study were: (1) to compare growth trajectories of speech recognition in noise for children with normal hearing (CNH) and children who are hard of hearing (CHH) and (2) to determine the effects of auditory access, vocabulary size, and working memory on growth trajectories of speech recognition in noise in CHH.

DESIGN: Participants included 290 children enrolled in a longitudinal study. Children received a comprehensive battery of measures annually, including speech recognition in noise, vocabulary, and working memory. We collected measures of unaided and aided hearing and daily hearing aid (HA) use to quantify aided auditory experience (i.e., HA dosage). We used a longitudinal regression framework to examine the trajectories of speech recognition in noise in CNH and CHH. To determine factors that were associated with growth trajectories for CHH, we used a longitudinal regression model in which the dependent variable was speech recognition in noise scores, and the independent variables were grade, maternal education level, age at confirmation of hearing loss, vocabulary scores, working memory scores, and HA dosage.

RESULTS: We found a significant effect of grade and hearing status. Older children and CNH showed stronger speech recognition in noise scores compared to younger children and CHH. The growth trajectories for both groups were parallel over time. For CHH, older age, stronger vocabulary skills, and greater average HA dosage supported speech recognition in noise.

CONCLUSION: The current study is among the first to compare developmental growth rates in speech recognition for CHH and CNH. CHH demonstrated persistent deficits in speech recognition in noise out to age 11, with no evidence of convergence or divergence between groups. These trends highlight the need to provide

support for children with all degrees of hearing loss in the academic setting as they transition into secondary grades. The results also elucidate factors that influence growth trajectories for speech recognition in noise for children; stronger vocabulary skills and higher HA dosage supported speech recognition in degraded situations. This knowledge helps us to develop a more comprehensive model of spoken word recognition in children.

Arch Dis Child. 2019 Nov;104(11):1056-1063. doi: 10.1136/archdischild-2019-316917. Epub 2019 May 11.

Academic, behavioural and quality of life outcomes of slight to mild hearing loss in late childhood: a population-based study.

Wang J, Quach J, Sung V, Carew P, Edwards B, Grobler A, Gold L, Wake M.

OBJECTIVE: To investigate the associations of hearing thresholds and slight to mild hearing loss with academic, behavioural and quality of life outcomes in children at a population level.

METHODS: *Design and participants:* children aged 11-12 years in the population-based cross-sectional Child Health CheckPoint study within the Longitudinal Study of Australian Children. *Audiometry:* mean hearing threshold across 1, 2 and 4 kHz (better and worse ear); slight/mild hearing loss (threshold of 16-40 decibels hearing loss (dB HL)). *Outcomes:* National Assessment Program - Literacy and Numeracy, language, teacher-reported learning, parent and teacher reported behaviour and self-reported quality of life. *Analysis:* linear regression quantified associations of hearing threshold/loss with outcomes.

RESULTS: Of 1483 children (mean age 11.5 years), 9.2% and 13.1% had slight/mild bilateral and unilateral hearing loss, respectively. Per SD increment in better ear threshold (5.7 dB HL), scores were worse on several academic outcomes (eg, reading 0.11 SD, 95% CI 0.05 to 0.16), parent-reported behaviour (0.06 SD, 95% CI 0.01 to 0.11) and physical (0.09 SD, 95% CI 0.04 to 0.14) and psychosocial (0.06 SD, 95% CI 0.01 to 0.11) Pediatric Quality of Life Inventory (PedsQL). Compared with normally hearing children, children with bilateral slight/mild losses scored 0.2-0.3 SDs lower in sentence repetition, teacher-reported learning and physical PedsQL but not other outcomes. Similar but attenuated patterns were seen in unilateral slight/mild losses.

CONCLUSIONS: Hearing thresholds and slight/mild hearing loss showed small but important associations with some child outcomes at 11-12 years. Justifying hearing screening or intervention at this age would require better understanding of its longitudinal and indirect effects, alongside effective management and appropriate early identification programmes.

Cochlear Implants Int. 2019 May;20(3):127-137. doi: 10.1080/14670100.2019.1573952. Epub 2019 Feb 6.

Effect of auditory status on visual emotion recognition in adolescents.

Warner-Czyz AD, Evans D, Turkstra L, Scheppele M, Song C, Evans JL.

Adolescents with severe to profound hearing loss who wear cochlear implants (CIs) experience significantly more peer problems compared to peers with typical hearing (TH). Differences in peer social dynamics may relate to perception not only of message content, but also message intent based on a speaker's emotion from visual (e.g. facial expressions) and auditory (e.g. prosody) cues. Pediatric CI users may experience greater difficulty with auditory emotion recognition due to an impoverished signal representation provided by the device, but the effect of auditory status on visual emotion recognition yields conflicting results.

OBJECTIVES: The current study examined accuracy and speed of visual emotion recognition in adolescents with CIs and peers with TH.

METHODS: Participants included 58 adolescents (10-18 years) stratified by auditory status: 34 CI users and 24 TH peers. Participants identified the intended emotion (i.e. happiness, sadness, anger, fear, disgust, and surprise) of static images of faces displayed on a computer screen.

RESULTS: No significant differences by auditory status emerged for response accuracy, response time to all trials, or response time to correct trials. Type of emotion significantly affected both accuracy and response time.

CONCLUSION: Adolescents with CIs show similar accuracy and response time in recognizing static facial expressions compared to TH peers. Future studies should explore the association between visual emotion recognition and social well-being to determine the relationship between emotion recognition and overall quality of life in adolescents with CIs.

Pediatr Infect Dis J. 2019 Jun;38(6S Suppl 1):S60-S63. doi: 10.1097/INF.0000000000002325.

Congenital Cytomegalovirus and Neonatal Herpes Simplex Virus Infections: To Treat or Not to Treat?

Whitley RJ.

Congenital cytomegalovirus infections are among the most common of the newborn in the developed world. These infections are the most common cause of sensorineural hearing loss. Studies utilizing ganciclovir and valganciclovir demonstrate improved hearing and Bailey Developmental scores. Because of the ease of

administration, valganciclovir is the recommended treatment of choice for 6 months. Therapy should be reserved for those babies with symptomatic disease; no data are available regarding the impact of treatment on those babies with asymptomatic disease.

Audiol Neurotol. 2019 Nov 1:1-12. doi: 10.1159/000503135. [Epub ahead of print]

BalanCI: Head-Referenced Cochlear Implant Stimulation Improves Balance in Children with Bilateral Cochleovestibular Loss.

Wolter NE, Gordon KA, Campos JL, Vilchez Madrigal LD, Pothier DD, Hughes CO, Papsin BC, Cushing SL.

INTRODUCTION: To determine the impact of a head-referenced cochlear implant (CI) stimulation system, BalanCI, on balance and postural control in children with bilateral cochleovestibular loss (BCVL) who use bilateral CI.

METHODS: Prospective, blinded case-control study. Balance and postural control testing occurred in two settings: (1) quiet clinical setting and (2) immersive realistic virtual environment (Challenging Environment Assessment Laboratory [CEAL], Toronto Rehabilitation Institute). Postural control was assessed in 16 and balance in 10 children with BCVL who use bilateral CI, along with 10 typically developing children. Children with neuromotor, cognitive, or visual deficits that would prevent them from performing the tests were excluded. Children wore the BalanCI, which is a head-mounted device that couples with their CIs through the audio port and provides head-referenced spatial information delivered via the intracochlear electrode array. Postural control was measured by center of pressure (COP) and time to fall using the WiiTM (Nintendo, WA, USA) Balance Board for feet and the BalanCI for head, during the administration of the Modified Clinical Test of Sensory Interaction in Balance (CTSIB-M). The COP of the head and feet were assessed for change by deviation, measured as root mean square around the COP (COP-RMS), rate of deviation (COP-RMS/duration), and rate of path length change from center (COP-velocity). Balance was assessed by the Bruininks-Oseretsky Test of Motor Proficiency 2, balance subtest (BOT-2), specifically, BOT-2 score as well as time to fall/fault.

RESULTS: In the virtual environment, children demonstrated more stable balance when using BalanCI as measured by an improvement in BOT-2 scores. In a quiet clinical setting, the use of BalanCI led to improved postural control as demonstrated by significant reductions in COP-RMS and COP-velocity. With the use of BalanCI, the number of falls/faults was significantly reduced and time to fall increased.

CONCLUSIONS: BalanCI is a simple and effective means of improving postural control and balance in children with BCVL who use bilateral CI. BalanCI could potentially improve the safety of these children, reduce the effort they expend maintaining balance and allow them to take part in more complex balance tasks where sensory information may be limited and/or noisy.

Laryngoscope. 2019 Oct 29. doi: 10.1002/lary.28329. [Epub ahead of print]

A novel tablet-based approach for hearing screening of the pediatric population, 516-patient study.

Xiao L, Zou B, Gao L, Weng M, Lando M, Smith AE, Barber W, Yao H.

OBJECTIVE: Assess a tablet hearing game as a screening instrument for pediatric hearing loss.

METHODS: All children age 3 to 13 presenting to the ENT clinic of a tertiary hospital clinic over a 3-month period were eligible for study. Five hundred sixteen were entered by completing the tablet screen with calibrated tablet/headphones. All had full standard audiometry or otoacoustic emission testing to assess hearing status. Tablet game data was analyzed to find the best correlation to the air conduction audiogram. The appropriate pass threshold of the tablet game was established and the statistical accuracy of the tablet game versus the air conduction audio was assessed.

RESULTS: The overall rate of hearing loss was 29.7% (153 subjects). Conductive hearing loss predominated and was present in 128 children. The tablet game pure tone average from 500- 4000 Hz correlated best with the air conduction audiogram, and was most predictive of hearing loss. Setting the pass level at 20 dB for the tablet screen prioritized detection of hearing loss, yielding a sensitivity of 91% and corresponding specificity of 73.5% for ages 4 and older. Specificity progressively improved with increasing age and was over 90% for all ages 7 and older.

CONCLUSION: Tablet game audiometry as a screening tool performs well in a controlled setting. Based on these results, it can be considered as a reliable screening method for school-age children and to monitor resolution of otitis media.

LEVEL OF EVIDENCE: 4, case series Laryngoscope, 2019.

Relationship research between auditory neuropathy spectrum disorder and exchange transfusion in neonates with severe hyperbilirubinemia.

[Xu J](#), [Weng M](#), [Li N](#), [Wu X](#), [Gao L](#), [Yao H](#), [Su S](#).

OBJECTIVE: To explore the effects of exchange transfusion on auditory neuropathy spectrum disorder (ANSD) in neonates with severe hyperbilirubinemia (SH).

METHODS: The clinical data of 2216 SH neonates who met the standard of exchange transfusion and 732 non severe-hyperbilirubinemia (NSH) neonates in the same period who did not require exchange transfusion in the neonatology department of Childrens' Hospital of Chongqing Medical University between January 2010 and December 2015 were retrospectively analyzed. In addition, the SH neonates were further divided into the exchange transfusion group and photography group. Hearing screening was conducted on all neonates using transiently evoked otoacoustic emission (TEOAE) and auto auditory brainstem response (AABR), and neonates who failed the above screening were performed diagnostic hearing test. And then neonates diagnosed with hearing disorder were followed up for 2-5 years.

RESULTS: The pass rates of hearing screening were 80.58%, 79.71% and 87.84% in the phototherapy group, exchange transfusion group and NSH group respectively, with a significant difference ($P < 0.05$). Hearing loss was diagnosed in 10.15%, 12.39% and 8.54% of neonates in the phototherapy group, exchange transfusion group and NSH group. After follow-up, ultimate incidence rates of ANSD were 11.96%, 11.57% and 2.4% respectively in the 3 groups, with a significant difference ($P < 0.05$).

CONCLUSIONS: SH is one of risk factors for ANSD. SH neonates have a lower incidence of ANSD in the exchange transfusion group than in the phototherapy group. Neonates who meet the standards of exchange transfusion adopt this therapy in early stage, which can quickly decrease bilirubin level and ultimately reduce incidence of ANSD.

Contribution of congenital cytomegalovirus (cCMV) to permanent hearing loss in a highly seropositive population: "The BraCHS study".

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

BACKGROUND: The exact contribution of congenital CMV infection (cCMV) to congenital and early-onset permanent hearing loss in highly seropositive populations is unknown.

OBJECTIVES: To determine the contribution of the cCMV to hearing impairment and to estimate the effectiveness of newborn hearing screening (HS) in identifying infants with CMV-related hearing loss.

STUDY DESIGN: A total of 11,900 infants born from a population with $\geq 97\%$ maternal seroprevalence were screened for cCMV and hearing loss. cCMV was confirmed by detection of CMV-DNA in saliva and urine < 3 weeks of age.

RESULTS: Overall, 68 (0.6%; CI95%:0.4-0.7) neonates were identified with cCMV. Of the 91 (0.8%) infants who failed HS, 24 (26.4%) were confirmed with hearing loss (HL) including 7 (29.2%; CI95%:17.2-59.3) with cCMV. Another infant with cCMV passed HS but was confirmed with HL at 21 days of age. Of the 62 infants with cCMV who underwent complete hearing evaluation, 8 (12.9%; CI95%: 6.7-23.4) had HL and most (7/8; 87.5%; CI95%:46.6-99.7) were identified by HS. The rate of permanent HL resulting from cCMV was 8 per 11,887 infants (0.7 per 1000 live births). The prevalence ratio of hearing loss among infants with cCMV compared to CMV-uninfected infants was 89.5 (CI95%:39.7-202.0). No late onset of CMV-related HL was detected during a median follow-up of 36 months.

CONCLUSIONS: cCMV is an important cause of permanent hearing impairment in childhood in all settings. Integrating targeted cCMV screening among infants who fail HS could be a reasonable cost-effective strategy to identify infants with early onset of cCMV-related HL.

Pediatric hearing screening in low-resource settings: Incorporation of video-otoscopy and an electronic medical record.

[Yancey KL](#), [Cheromei LJ](#), [Muhandu J](#), [Reppart J](#), [Netterville JL](#), [Jayawardena ADL](#).

OBJECTIVE: To develop a sustainable, comprehensive, intervention-based approach to pediatric hearing care in low- and middle income countries (LMICs) where global hearing loss is most heavily concentrated.

METHODS: Community health workers (CHWs) in Kilifi county, Kenya were trained to conduct hearing screening and video-otoscopy via a unified, smartphone-based platform using mobile electronic medical record (EMR)

generation for children diagnosed with hearing loss or other pathology. Among at-risk students pre-selected by their teachers, the frequency of hearing loss and pathology in children with and without hearing loss was measured.

RESULTS: Of the 155 screened, 16 (10%) children were found to have hearing loss. 12 (5.9%) children with normal hearing had the following pathology: perforation (N=5 ears), effusion (N=9), retraction (N=6), and infections (N=7). CHWs were also adept at EMR creation without significant delay in workflow. Out of all those screened, 28 (18%) children were found to have hearing loss or other pathology and were referred to follow up. All 28 of 28 children referred were successfully entered into the EMR.

CONCLUSIONS: CHWs with little to no prior medical experience can provide a much needed public health service - hearing screening in LMICs where access to health care is limited. The incorporation of video-otoscopy provides a more comprehensive approach to hearing care by not only helping identify etiologies of existing hearing disability but also conditions that predispose to future hearing loss. It can easily be performed in conjunction with hearing screenings via the use of a unified, mobile platform. The addition of EMR supports follow-up and allows remote consultation.

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J Otolaryngol Head Neck Surg. 2019 May 22;48(1):22. doi: 10.1186/s40463-019-0341-z.

Subjective and objective vestibular changes that occur following paediatric cochlear implantation: systematic review and meta-analysis.

Yong M, Young E, Lea J, Foggini H, Zaia E, Kozak FK, Westerberg BD.

OBJECTIVE: Cochlear implantation can result in post-operative vestibular dysfunction of unknown clinical significance. The objective of this study was to characterize the presence, magnitude, and clinical significance of vestibular dysfunction that occurs after pediatric cochlear implantation.

DATA SOURCES: The databases Embase, Medline (OvidSP), and PubMed were used. Only articles published in English were included. Grey literature and unpublished sources were also reviewed.

STUDY SELECTION: Articles published from 1980 until the present which documented pre-operative and post-operative vestibular testing on children under the age of 18 were used.

DATA EXTRACTION: Parameters that were assessed included number of patients, pre- and post-operative vestibular-evoked myogenic potentials (VEMPs), head impulse testing (HIT), calorics, and posturography, timing of pre- and post-operative testing, symptomatology, and other demographic data such as etiology of the hearing loss.

DATA SYNTHESIS: Ten articles were included. Relative risk values evaluating the effect of cochlear implantation on vestibular function were calculated for VEMPs and caloric testing due to the availability of published data. I^2 values were calculated and 95% confidence intervals were reported. Separate analyses were conducted for each individual study and a pooled analysis was conducted to yield an overall relative risk. Assessment on risk of bias in individual studies and overall was performed.

CONCLUSION: Pediatric cochlear implantation is associated with a statistically significant decrease in VEMP responses post-operatively (RR 1.8, $p < 0.001$, I^2 91.86, 95%CI 1.57-2.02). Similar results are not seen in caloric testing. Insufficient data is available for analysis of HIT and posturography. Further studies are necessary to determine the effect of cochlear implantation on objective vestibular measures post-operatively and whether any changes seen are clinically relevant in this population.

Int J Pediatr Otorhinolaryngol. 2019 Aug;123:187-190. doi: 10.1016/j.ijporl.2019.05.019. Epub 2019 May 21.

Newborn hearing screening results of refugees living in our city and the factors affecting the results.

Yücel A, Alataş N, Yücel H, Güllüev M, Özsöz E, Uğur C.

OBJECTIVES: To investigate the changes in the risk factors affecting the results of the Newborn Hearing Screening (NHS) and the hearing test results of the Syrian refugees in our city.

METHODS: Syrian and Turkish newborns, born in our hospital between 01.01.2016 and 31.12.2017 and referred to our hospital from environmental hospitals for NHS, were included in this study. NHS results and risk factors were analyzed.

RESULTS: 786 Syrian and 7230 Turkish newborns were included in this study. 53 (6,74%) infants referred in both ears, 26 (3,30%) infants in the one ear. There was a significant relationship between the presence of hearing loss and the history of intensive care unit admittance, presence and absence of low birth weight and neonatal icterus at Syrian newborns. In the same period, 20 (0,3%) Turkish infants referred bilaterally and 45 (0,6%) newborns unilaterally (25 right ear, 20 left ear). There was a significant difference between Turkish and Syrian newborns in terms of very low and low birth weight and intensive care unit admittance.

CONCLUSIONS: The rate of hearing loss in Syrian refugee patients is quite high. Pregnant refugee women who are forced to migrate because of war face many risk factors and these people need to be included into the newborn hearing screening programs in the country where they took refuge in.

Pediatrics. 2019 Oct;144(4). pii: e20190811. doi: 10.1542/peds.2019-0811.

Prevalence and Trends of Developmental Disabilities among Children in the United States: 2009-2017.

Zablotsky B, Black LI, Maenner MJ, Schieve LA, Danielson ML, Bitsko RH, Blumberg SJ, Kogan MD, Boyle CA.

OBJECTIVES: To study the national prevalence of 10 developmental disabilities in US children aged 3 to 17 years and explore changes over time by associated demographic and socioeconomic characteristics, using the National Health Interview Survey.

METHODS: Data come from the 2009 to 2017 National Health Interview Survey, a nationally representative survey of the civilian noninstitutionalized population. Parents reported physician or other health care professional diagnoses of attention-deficit/hyperactivity disorder; autism spectrum disorder; blindness; cerebral palsy; moderate to profound hearing loss; learning disability; intellectual disability; seizures; stuttering or stammering; and other developmental delays. Weighted percentages for each of the selected developmental disabilities and any developmental disability were calculated and stratified by demographic and socioeconomic characteristics.

RESULTS: From 2009 to 2011 and 2015 to 2017, there were overall significant increases in the prevalence of any developmental disability (16.2%-17.8%, $P < .001$), attention-deficit/hyperactivity disorder (8.5%-9.5%, $P < .01$), autism spectrum disorder (1.1%-2.5%, $P < .001$), and intellectual disability (0.9%-1.2%, $P < .05$), but a significant decrease for any other developmental delay (4.7%-4.1%, $P < .05$). The prevalence of any developmental disability increased among boys, older children, non-Hispanic white and Hispanic children, children with private insurance only, children with birth weight ≥ 2500 g, and children living in urban areas and with less-educated mothers.

CONCLUSIONS: The prevalence of developmental disability among US children aged 3 to 17 years increased between 2009 and 2017. Changes by demographic and socioeconomic subgroups may be related to improvements in awareness and access to health care.

JAMA Otolaryngol Head Neck Surg. 2019 Oct 10. doi: 10.1001/jamaoto.2019.2877. [Epub ahead of print]

Identification of Potential Barriers to Timely Access to Pediatric Hearing Aids.

Zhang L, Links AR, Boss EF, White A, Walsh J.

IMPORTANCE: Despite various barriers identified to early pediatric access to cochlear implantation, barriers to timely access to pediatric hearing aids are not well characterized.

OBJECTIVE: To identify socioeconomic, demographic, and clinical factors that may be associated with pediatric access to hearing aids.

DESIGN, SETTING, AND PARTICIPANTS: This retrospective cohort study included 90 patients aged 1 to 15 years who were referred for auditory brainstem response (ABR) testing and evaluation for hearing aids at a single tertiary care academic medical center from March 2004 to July 2018. Children who did not receive both ABR testing and hearing aids at the same center were excluded from analysis.

MAIN OUTCOMES AND MEASURES: Associations of insurance type (private vs public), race/ethnicity (white vs other), primary language (English vs other), cause of hearing loss (complex vs not complex), zip code, hearing aid manufacturer, and severity of hearing loss (in decibels) with the duration of intervals from newborn hearing screening to ABR testing, from ABR testing to ordering of hearing aids, and from ABR testing to dispensing of hearing aids.

RESULTS: Of the 90 patients, mean (SD) age was 5.6 (3.7) years, 56% were female, and 77 (86%) were non-Hispanic. Results of χ^2 tests indicated significant associations existed between public insurance and race/ethnicity and between public insurance and primary language other than English. Variables associated with the interval from newborn hearing screening to ABR testing included insurance type (mean difference, 7.4 months; 95% CI, 2.6-12.2 months) and race/ethnicity (mean difference, 6.9 months; 95% CI, 2.7-11.1 months). Increased delays between birth and a child's first ABR test were associated with public insurance (mean difference, 6.0 months; 95% CI, 1.8-10.2 months) and race/ethnicity other than white (mean difference, 6.0 months; 95% CI, 2.3-9.7 months). The mean time from birth to initial ABR testing was a mean of 6 months longer for patients from non-English-speaking families than for those from English-speaking families (mean [SD] interval, 14.9 [16.3] months vs 9.0 [8.5] months), although the difference was not statistically significant. Severity of hearing loss was associated with a decrease in the interval from ABR testing to ordering of hearing aids after accounting for other potential barriers (odds ratio, 0.6; 95% CI, 0.4-0.9). Zip code and complexity of the child's medical condition did not appear to be associated with timely access to pediatric hearing aids.

CONCLUSIONS AND RELEVANCE: This study's findings suggest that insurance type, race/ethnicity, and primary language may be barriers associated with pediatric access to hearing aids, with the greatest difference observed in time to initial ABR testing. Clinical severity of hearing loss appeared to be associated with a significant decrease in time from ABR testing to ordering of hearing aids. Greater efforts to assist parents with ABR testing and coordination of follow-up may help improve access for other at-risk children.

Biosci Trends. 2019 Jul 22;13(3):261-266. doi: 10.5582/bst.2019.01142. Epub 2019 Jun 25.

[Analysis of mutations in the FOXI1 and KCNJ10 genes in infants with a single-allele SLC26A4 mutation.](#)

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

The current study investigated how the FOXI1 and KCNJ10 genes were affected in infants with a single-allele mutation in the SLC26A4 gene, and it determined the audiological phenotypes of infants with double heterozygous mutations (DHMs) in the three genes. Subjects were 562 infants with a single-allele SLC26A4 mutation detected during neonatal deafness genetic screening; the infants were seen as outpatients by Otology at Beijing Tongren Hospital. All subjects underwent SLC26A4 sequencing. Twenty infants had a second-allele variant while the remaining 542 had an SLC26A4 single-allele mutation. Infants also underwent FOXI1 and KCNJ10 sequencing. All patients with double heterozygous mutations in the aforementioned genes underwent an audiological evaluation and a limited imaging study; variants and audiological phenotypes were analyzed. Of 562 patients, 20 had SLC26A4 bi-allelic mutations; 8 carried single mutations in both SLC26A4 and KCNJ10. No pathogenic mutations in the FOXI1 gene were found. Four missense mutations in KCNJ10 were detected, including c.812G>A, c.800A>G, c.53G>A, and c.1042C>T. Eight individuals with a DHMs all passed universal newborn hearing screening, and all were found to have normal hearing. These data suggest that individuals with an SLC26A4 single-allele mutation, combined with FOXI1 or KCNJ10 gene mutations, do not suffer hearing loss during infancy, though this finding is worthy of further follow-up and in-depth discussion.

Am J Otolaryngol. 2019 Jul - Aug;40(4):573-576. doi: 10.1016/j.amjoto.2019.05.011. Epub 2019 May 14.

[Impact of metabolic syndrome on recovery of idiopathic sudden sensorineural hearing loss.](#)

[Zhou Y](#), [Qiu S](#), [Liu D](#).

PURPOSE: Metabolic syndrome (MetS) was reported to a risk factor of developing idiopathic sudden sensorineural hearing loss (ISSNHL), but limited data exist on its effect on the recovery. The purpose of this study was to evaluate the impact of (MetS) and its components on recovery of patients with ISSNHL.

MATERIAL AND METHODS: 228 ISSNHL patients were divided into MetS group and Non-MetS group according to the diagnostic criteria of MetS, and demographic and clinical characteristics and hearing recovery were reviewed between two groups.

RESULTS: In total, 86 (37.7%) patients in MetS group, and 142 (62.3%) patients in Non-MetS group. The rate of hypertension, diabetes mellitus, low HDL-C, high TG and obesity were significantly higher in the MetS group than those in the Non-MetS group ($P < 0.05$). The complete recovery rate and partial recovery rate were significantly lower in the MetS group than those in the Non-MetS group. According to the multivariate analysis, MetS was significantly associated with a poor prognosis; high initial hearing threshold and presence of diabetes mellitus were correlated with a poor prognosis ($P < 0.05$).

CONCLUSIONS: These results suggest that MetS has a negative impact on the hearing recovery of ISSNHL. High initial hearing threshold and diabetes mellitus were indicators of a poor prognosis of ISSNHL.

Int J Pediatr Otorhinolaryngol. 2019 Nov;126:109630. doi: 10.1016/j.ijporl.2019.109630. Epub 2019 Aug 7.

[Suspension array-based deafness genetic screening in 53,033 Chinese newborns identifies high prevalence of 109G>A in GJB2.](#)

[Zou Y](#), [Dai QQ](#), [Tao WJ](#), [Wen XL](#), [Feng DF](#), [Deng H](#), [Zhou WP](#), [Li M](#), [Zhang L](#).

OBJECTIVES: More than 50% of congenital hearing loss is attributed to genetic factors. Data of gene mutation associated with hearing loss from large population studies in Chinese population are scarce. In this study, we conducted a comprehensive newborn genetic screening in China to establish the carrier frequency and mutation spectrum of deafness-associated genes.

METHODS: A total of 53,033 newborns were screened for hearing defects associated mutations. Twenty hot spot mutations in GJB2, GJB3, SLC26A4 and mitochondria12S rRNA were examined using suspension array analysis.

RESULTS: 14,185 newborns (26.75%) were identified with at least one mutated allele. 872 (1.64%) neonates carried homozygous mutations including 112 (0.21%) mitochondrial DNA homoplasmy, 228 (0.43%) were compound heterozygotes, and 11,985 (22.59%) were heterozygotes including 11 (0.02%) mitochondrial DNA heteroplasmy. Top five mutations included 109 G>A, 235 delC, 299-300 delAT in GJB2, IVS7-2 A>G in SLC26A4 and 1555 A>G in mitochondria12S rRNA. Notably, a total of 10,995 neonates (20.73%) carried 109 G>A in GJB2. Moreover, the allele frequencies of 109 G>A were detected 11.61% in Guangdong, 10.44% in Sichuan and 2.88%

in Shandong, respectively, a significant difference in prevalence among these geographic regions ($p < 0.01$). In addition, the high frequency of 109 G>A in GJB2 was confirmed by a TaqMan probe-based qPCR assay. Very recently, the ClinGen Hearing Loss Expert Panel reached a consensus and confirmed its pathogenic role in hearing impairment.

CONCLUSION: We delineated the mutation profile of common deafness-causing genes in the Chinese population and highlighted the high prevalence of 109 G>A pathogenic mutation. Our study may facilitate early diagnosis/ intervention and genetic counseling for hearing impairment in clinical practice.



Wearing Hearing Aids improves language skills IF WORN 10 HOURS OR MORE EACH DAY.



Put hearing aids on -----> When awake

In the car <-----



-----> At play



- ☒ Sunday
- ☒ Monday
- ☒ Tuesday
- ☒ Wednesday
- ☒ Thursday
- ☒ Friday
- ☒ Saturday



Every day

Talk to your audiologist about how best to meet the needs of your family

Reference: Tomblin, J.B., Harrison, M., Ambrose, S.E., Walker, E.A., Oleson, J.J. & Moeller, M.P. (2015). Language outcomes in young children with mild to severe hearing loss. *Ear and Hearing*, 36, 76S-91S. doi: 10.1097/AUD.0000000000000219