The Journal of Early Hearing Detection and Intervention

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Others' Publications About EHDI: October 2018 through April 2019

The J*ournal 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 (EHDI) systems.

Although JEHDI is the only journal that focuses exclusively on improving EHDI 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 EHDI programs, we provide titles and abstracts of recent publications that are relevant to improving EHDI programs. Titles of all articles are hyperlinked to the source.

EHDI continues to be a global phenomenon. Of the 118 abstracts of articles included in the following abstracts, almost 50% are from authors in low and middle income countries. Many of the abstracts listed below focus on the basic components of EHDI systems (e.g., screening, diagnosis, early intervention), suggesting that there are still areas in the basic EHDI system that need to be improved. Other publications report studies about how to best incorporate detection of hearing loss in screening programs designed to detect conditions such as congenital cytomegalovirus and newborn genetic screening. There are also a number of studies about what causes hearing loss. For example Brennan-Jones et al. did a comprehensive review showing that children treated for childhood cancer using platinum analogues had more hearing loss than other children. The topic of childhood cancer treatment and hearing loss was also addressed by Clemens et al., Robertson et al., and Weiss et al. A number of studies from around the world also examined comorbidities of childhood hearing loss. For example, there were five articles that examined the incidence of childhood hearing loss among children diagnosed with sickle cell disease (Farrell et al., Lago et al., Rissatto-Lago et.al, Schopper et al, and Towerman et al.) and De Schrijver et al. looked at the incidence of hearing loss among children with Down syndrome and concluded that it is not as prevalent as many people think. Knowing more about the conditions that cause childhood hearing loss and what other groups of children are affected with hearing loss, will help to improve the efficiency and effectiveness of EHDI systems.

Below are examples of other interesting findings of recently reported studies from around the world.

- Bartlett et al., based on a newborn screening program for congenital cytomegalovirus (CMV) that identified 214 cases of symptomatic and 88 cases of asymptomatic CMV, concluded that universal newborn CMV screening should be considered for implementation.
- Cedars et al., in a study with 3,257 children concluded that hearing screening using a combination of conditioned play audiometry and otoacoustic emissions testing in a preschool setting reduced referral rates, increased identification of hearing loss, reduced outcome disparities, and improved follow-up rates.
- Fitzpatrick et al. studied a total of 120 children (38 with unilateral hearing loss, 31 with bilateral mild hearing loss, and 51 with normal hearing) and concluded that even when they are identified during the first few months of life, children with unilateral hearing loss tend to lag behind their peers in receptive and expressive language development.
- Puia-Dumitrescu et al., in a study of gentamicin use in neonatal intensive care units that involved over 80,000 children concluded that use of gentamicin, regardless of dose and length of treatment was not associated with increased odds of failing the newborn hearing screen.
- Ramkumar et al., concluded that community-based pediatric screening in rural parts of India could be done more effectively using a telepractice model for diagnostic follow-up with auditory brainstem response compared to an in-person evaluation at a tertiary care hospital with auditory brainstem response testing.
- Sözen et al., in a study of the effect of a national pneumococcal vaccination program done in Turkey found that the incidence of meningitis-induced hearing loss had been reduced by more than ten-fold since the implementation of the program.
- Walker et al. found that only about one-third of preschool-aged children who are hard of hearing have access to a remote microphone system for home use, and about one-half for school use. For those children who have access to a remote microphone system, average use was only about 1–2 hours at home and 2–4 hours in school.

Abstracts of many more articles with results that are important for continuing to improve EHDI systems are listed below.

Abulebda K, Patel VJ, Ahmed SS, Tori AJ, Lutfi R, Abu-Sultaneh S.
Comparison between chloral hydrate and propofol-ketamine as sedation regimens for pediatric auditory brainstem response testing.
Braz J Otorhinolaryngol. 2019 Jan - Feb;85(1):32-36. doi: 10.1016/j.bjorl.2017.10.003. Epub 2017 Oct 28.

INTRODUCTION: The use of diagnostic auditory brainstem response testing under sedation is currently the "gold standard" in infants and young children who are not developmentally capable of completing the test.
OBJECTIVE: The aim of the study is to compare a propofol-ketamine regimen to an oral chloral hydrate regimen for sedating children undergoing auditory brainstem response testing.
METHODS: Patients between 4 months and 6 years who required sedation for auditory brainstem response testing were included in this retrospective study. Drugs doses, adverse effects, sedation times, and the effectiveness of the sedative regimens were reviewed.
RESULTS: 73 patients underwent oral chloral hydrate sedation, while 117 received propofol-ketamine sedation. 12% of the patients in the chloral hydrate group failed to achieve desired sedation level. The average procedure, recovery and total nursing times were significantly lower in the propofol-ketamine group. Propofol-ketamine group experienced higher incidence of transient hypoxemia.
CONCLUSION: Both sedation regimens can be successfully used for sedating children undergoing auditory brainstem response testing. While deep sedation using propofol-ketamine regimen offers more efficiency than moderate sedation using chloral hydrate, it does carry a higher incidence of transient hypoxemia, which warrants the use of a highly skilled team trained in pediatric cardio-respiratory monitoring and airway management.

Aloqaili Y, Arafat AS, Almarzoug A, Alalula LS, Hakami A, Almalki M, Alhuwaimel L.
Knowledge about cochlear implantation: A parental perspective.
Cochlear Implants Int. 2018 Nov 22:1-6. doi: 10.1080/14670100.2018.1548076.

OBJECTIVES: Cochlear implantation (CI) is used for children with severe to profound hearing loss who show little or no improvement using hearing aids. This study explored parental knowledge of their children's CI.
METHODS: A cross-sectional study involving the parents of 115 pediatric CI patients was conducted at King Abdullah Specialized Children's Hospital in Riyadh, Saudi Arabia. Parents were interviewed by telephone using a 50-question validated questionnaire.
RESULTS: Most parents of children with CI reported being comfortable in using the internet (68.7%) and social media (40.9%) to obtain information regarding CI. Although most parents of children with CI relied on health professionals and websites as their main sources of information, they were also able to obtain necessary information at meetings for CI patients and health professionals. Parents of children with CI felt they had sufficient information regarding the impact of hearing loss (78%) and CI (71%) on speech understanding and language development; however, they had insufficient information regarding criteria for CI candidacy, available brands of CI devices, and the advantages and disadvantages of each.
CONCLUSION: Parents reported that health professionals were the ideal source of information regarding hearing loss and CI. Moreover, our study showed that parents should learn more about cochlear implant devices, the post-implantation process, and candidacy criteria.

Ameyaw GA, Ribera J, Anim-Sampong S.
Interregional Newborn Hearing Screening via Telehealth in Ghana.
J Am Acad Audiol. 2019 Mar;30(3):178-186. doi: 10.3766/jaaa.17059. Epub 2018 Feb 7.

BACKGROUND: Newborn hearing screening is a vital aspect of the Early Hearing Detection and Intervention program, aimed at detecting hearing loss in children for prompt treatment. In Ghana, this kind of pediatric hearing service is available at only one health care facility located in the Greater Accra Region. The current practice in effect has virtually cut-off infants in the other regions from accessing hearing screening and other pediatric audiological services. This has prompted a study into alternative methodologies to expand the reach of such services in Ghana. The present study was designed to assess the feasibility of using telehealth to deliver newborn hearing screening across Ghana.
PURPOSE: To assess the feasibility of using telehealth to extend newborn hearing screening services across the ten regions of Ghana.
RESEARCH DESIGN: A correlational study was designed to determine the extent of association between test results of telehealth and the conventional on-site methods (COMs) for conducting newborn hearing screening. The design also allowed for testing duration between the two methods to be compared.
STUDY SAMPLE: Fifty infants from the Brong-Ahafo Regional Hospital (BARH) were enrolled. The infants aged between 2 and 90 days were selected through convenience sampling. There were 30 males and 20 females.

PROCEDURE: Newborn hearing screening using distortion product otoacoustic emissions were performed via telehealth. By adopting the synchronous telehealth model, an audiologist located at the Korle-Bu Teaching Hospital conducted real-time hearing screening tests over the internet on infants who were at the BARH. The former and latter hospitals are located in the Greater Accra and the Brong-Ahafo Regions, respectively. As a control, similar hearing screening tests were conducted on the same infants at BARH using the conventional face-to-face on-site hearing screening method.
DATA COLLECTION AND ANALYSIS: The test results and testing duration of the telehealth method and the conventional on-site approach were compared and subjected to statistical analysis. Here, the Spearman's correlation coefficient (r_s) was used to determine the level of correlation between the test results, whereas the paired t-test statistic was used to test the level of significance between the testing duration of the two methods.
RESULTS: Analysis of the test results showed a significantly high positive correlation between the telehealth and the COMs (r_s = 0.778, 0.878, 0.857, 0.823, $p < 0.05$ @ 2.0, 3.0, 4.0, and 5.0 kHz respectively). Also, the difference in testing duration of the two methods was not statistically significant [$t_{(99)} = 1.309$, $p > 0.05$]. The mean testing duration (in seconds) of telehealth was 27.287 (standard deviation = 27.373) and that of the COM was 24.689 (standard deviation = 27.169).
CONCLUSION: The study showed the feasibility of establishing an interregional network of newborn hearing screening services across Ghana using telehealth. It is more efficient to deploy telehealth for pediatric hearing services than to have patients travel many hours to the Greater Accra Region for similar services. Poor road network, high transportation costs, and bad weather conditions are a few of the reasons for avoiding long distance travel in Ghana.

Bartlett AW, Hall BM, Palasanthiran P, McMullan B, Shand AW, Rawlinson WD.
Recognition, treatment, and sequelae of congenital cytomegalovirus in Australia: An observational study.
J Clin Virol. 2018 Nov;108:121-125. doi: 10.1016/j.jcv.2018.09.017. Epub 2018 Sep 27.

BACKGROUND AND OBJECTIVES: Australian national surveillance data was used to assess recognition, sequelae, and antiviral therapy for congenital cytomegalovirus (CMV) cases.
STUDY DESIGN: Data from congenital CMV cases reported through the Australian Paediatric Surveillance Unit born January 1999 to December 2016 were described and Chi-square tests used to characterise trends and associations in case reporting, maternal CMV serology testing, and antiviral therapy. Descriptive analyses for hearing loss and developmental delay were reported for cases born ≥2004, following introduction of universal neonatal hearing screening.
RESULTS: There were 302 congenital CMV cases (214 symptomatic, 88 asymptomatic). Congenital CMV was suspected in 70.6% by 30 days of age, with no differences across birth cohorts. Maternal CMV serology testing was associated with maternal illness during pregnancy but not birth cohort. There was increasing antiviral use for symptomatic cases, being used in 14% born 1999-2004, 19.6% born 2005-2010, and 44.4% born 2011-2016 ($p < 0.001$). For those born ≥2004, hearing loss was reported in 42.1% of symptomatic and 26.6% of asymptomatic cases; while developmental delay was reported in 16.9% of symptomatic and 1.3% of asymptomatic cases.
CONCLUSION: There appears to be under-reporting and under-recognition of congenital CMV despite increasing use of antiviral therapy. Universal newborn CMV screening should be considered to facilitate follow-up of affected children and targeted linkage into hearing and developmental services, and to provide population-level infant CMV epidemiology to support research and evaluation of antiviral and adjunctive therapies.

Beaula Vincy VK, Seethapathy J, Boominathan P.
Parental anxiety towards 'refer' results in newborn hearing screening (NHS) in south India: A hospital based study.
Int J Pediatr Otorhinolaryngol. 2019 Jan;116:25-29. doi: 10.1016/j.ijporl.2018.10.021. Epub 2018 Oct 13.

BACKGROUND: Newborn Hearing Screening (NHS) aims at the early detection and intervention for children with congenital hearing loss. In developing countries like India, not all hospitals and birthing suites are equipped with NHS unit but there are few well established and emerging NHS programs that are operating in many parts of India. However, these screening procedures sometimes result in high false positive rates.
METHOD: This was a prospective cross sectional study. A total of 140 parents (parents of 70 well babies & parents of 70 NICU babies) of babies who underwent NHS between June, 2014 and December, 2014 at Sri Ramachandra Medical Centre (SRMC) were recruited for the study. Written parent consent was obtained prior to hearing screening. Parents of infants were counselled regarding the benefits of hearing screening, procedure of the screening test and need for follow-up testing if the neonate did not pass the screening test. Majority of the parents of infants (75%) were college graduates, 13% and 12% of parents had an educational level of high school and middle school respectively. Based on the Kuppuswamy's socioeconomic status scale, 69% of the parents were from upper middle class, 26% were from upper class and 5% were from lower middle class.
RESULTS: The present study aimed to identify parental anxiety towards 'refer' results of infants in the initial

NHS. Mean and standard deviation were used to find the state and trait anxiety levels in parents of each group. **CONCLUSION:** Refer' results in NHS lead to increased anxiety levels in parents of both well babies, and NICU babies. The increased anxiety levels may have greater impact on the parent's emotional status. Educating parents about screening procedures, possible causes for 'refer' results prior to screening, and also efforts to minimize false positive results in NHS can minimize unwanted anxiety in parents. At the same time, it is important that 'refer' results should be clearly explained and not minimized to ensure effective follow up. The audiologist dealing with NHS should take all attempts to alleviate anxiety in parents through public education, counseling and assertion.

Beswick R, David M, Higashi H, Thomas D, Nourse C, Koh G, Koorts P, Jardine L, Clark JE.
Integration of congenital cytomegalovirus screening within a newborn hearing screening programme.
J Paediatr Child Health. 2019 Mar 27. doi: 10.1111/jpc.14428. [Epub ahead of print]

AIM: Targeted screening by a salivary cytomegalovirus (CMV) polymerase chain reaction (PCR) of infants who 'refer' on their newborn hearing screen has been suggested as an easy, reliable and cost-effective approach to identify and treat babies with congenital CMV (cCMV) to improve hearing outcomes. This study aimed to investigate the feasibility and cost-effectiveness of introducing targeted salivary cCMV testing into a newborn hearing screening programme.
METHODS: The study included three tertiary maternity hospitals in Queensland, Australia between August 2014 and April 2016. Infants who 'referred' on the newborn hearing screen were offered a salivary swab for CMV PCR at the point of referral to audiology. Swabs were routinely processed and tested for CMV DNA by real-time quantitative PCR. Parents of babies with a positive CMV PCR were notified, and the babies were medically assessed and, where appropriate, were offered treatment (oral valganciclovir).
RESULTS: Of eligible infants, the parents of 83.0% (234/283) consented to the cCMV screen. Of these, 96.6% returned a negative result (226/234), and 3.4% (8/234) returned a positive result (three true positive; five false positive). The prevalence of cCMV for infants with confirmed hearing loss was 3.64% (P = 2/55; confidence interval = 0.44-12.53%). The cost comparison suggests the cost implementation of cCMV screening (and subsequent potential treatment benefits and management over time), compared to non-screening (and subsequent management), to be negligible.
CONCLUSION: Incorporating cCMV testing into Universal Newborn Hearing Screening within Queensland is realistic and achievable, both practically and financially.

Bezdjian A, Smith RA, Thomeer HGXM, Willie BM, Daniel SJ.
A Systematic Review on Factors Associated With Percutaneous Bone Anchored Hearing Implants Loss.
Otol Neurotol. 2018 Dec;39(10):e897-e906. doi:10.1097/MAO.0000000000002041.

OBJECTIVE: To investigate factors associated with percutaneous bone anchored hearing implant (BAHI) loss.
DATA SOURCES: Africa-Wide, Biosis, Cochrane, Embase, Global Health, LILACs, Medline, Pubmed, and Web of Science electronic databases.
STUDY SELECTION: All studies reporting on adult and/or pediatric patients with a BAHI loss were identified. Retrieved articles were screened using predefined inclusion criteria. Eligible studies underwent critical appraisal for directness of evidence and risk of bias. Studies that successfully passed critical appraisal were included for data extraction.
DATA EXTRACTION: Extracted data included study characteristics (study design, number of total implants and implant losses, follow-up), patient characteristics (sex, age, comorbidities, previous therapies), and information regarding BAHI loss (etiology of loss, timing of occurrence).
DATA SYNTHESIS: From the 5,151 articles identified at the initial search, 847 remained after title and abstract screening. After full text review, 96 articles were eligible. Fifty-one articles passed quality assessment, however, due to overlapping study population, 48 articles reporting on 34 separate populations were chosen for data extraction. Three hundred one implant losses occurred out of 4,116 implants placed, resulting in an overall implant loss occurrence rate of 7.3%. Failed osseointegration was responsible for most implant losses (74.2%), followed by fixture trauma (25.7%). Most losses due to failed osseointegration occurred within 6 months of the implantation. BAHI implant loss occurred more frequently in pediatric patients ($p<0.005$).
CONCLUSION: The current systematic review identified factors associated with BAHI loss. These factors should be considered when assessing patients' candidacy and when investigating reasons for impeded implant stability and loss.

Blankenship CM, Hunter LL, Keefe DH, Feeney MP, Brown DK, McCune A, Fitzpatrick DF, Lin L.
Optimizing Clinical Interpretation of Distortion Product Otoacoustic Emissions in Infants.
Ear Hear. 2018 Nov/Dec;39(6):1075-1090. doi: 10.1097/AUD.0000000000000562.

OBJECTIVES: The purpose of this study was to analyze distortion product otoacoustic emission (DPOAE) level and signal to noise ratio in a group of infants from birth to 4 months of age to optimize prediction of hearing

status. DPOAEs from infants with normal hearing (NH) and hearing loss (HL) were used to predict the presence of conductive HL (CHL), sensorineural HL (SNHL), and mixed HL (MHL). Wideband ambient absorbance was also measured and compared among the HL types.

DESIGN: This is a prospective, longitudinal study of 279 infants with verified NH and HL, including conductive, sensorineural, and mixed types that were enrolled from a well-baby nursery and two neonatal intensive care units in Cincinnati, Ohio. At approximately 1 month of age, DPOAEs (1-8 kHz), wideband absorbance (0.25-8 kHz), and air and bone conduction diagnostic tone burst auditory brainstem response (0.5-4 kHz) thresholds were measured. Hearing status was verified at approximately 9 months of age with visual reinforcement audiometry (0.5-4 kHz). Auditory brainstem response air conduction thresholds were used to assign infants to an NH or HL group, and the efficacy of DPOAE data to classify ears as NH or HL was analyzed using receiver operating characteristic (ROC) curves. Two summary statistics of the ROC curve were calculated: the area under the ROC curve and the point of symmetry on the curve at which the sensitivity and specificity were equal. DPOAE level and signal to noise ratio cutoff values were defined at each frequency as the symmetry point on their respective ROC curve, and DPOAE results were combined across frequency in a multifrequency analysis to predict the presence of HL.

RESULTS: Single-frequency test performance of DPOAEs was best at mid to high frequencies (3-8 kHz) with intermediate performance at 1.5 and 2 kHz and chance performance at 1 kHz. Infants with a conductive component to their HL (CHL and MHL combined) displayed significantly lower ambient absorbance values than the NH group. No differences in ambient absorbance were found between the NH and SNHL groups. Multifrequency analysis resulted in the best prediction of HL for the SNHL/MHL group with poorer sensitivity values when infants with CHL were included.

CONCLUSIONS: Clinical interpretation of DPOAEs in infants can be improved by using age-appropriate normative ranges and optimized cutoff values. DPOAE interpretation is most predictive at higher F2 test frequencies in young infants (2-8 kHz) due to poor test performance at 1 to 1.5 kHz. Multifrequency rules can be used to improve sensitivity while balancing specificity. Last, a sensitive middle ear measure such as wideband absorbance should be included in the test battery to assess possibility of a conductive component to the HL.

Bouillot L1, Vercherat , Durand C.

Implementing universal newborn hearing screening in the French Rhône-Alpes region. State of affairs in 2016 and the 1st half of 2017.

Int J Pediatr Otorhinolaryngol. 2019 Feb;117:30-36. doi: 10.1016/j.ijporl.2018.11.011. Epub 2018 Nov 10.

INTRODUCTION: Universal newborn hearing screening (UNHS) started as public health policy in 2015 in the French Rhône-Alpes region, aiming to screen for unilateral and bilateral hearing loss. After a first and second screening (retest) in the maternity hospital, the diagnostic process occurred at a limited number of specialist centers. A deferred preliminary screening (T3) was proposed before the age of 1 month. The aims of this study were to assess implementation of the program, impact of T3, and present the incidence of hearing loss in this population.

MATERIALS AND METHODS: The retrospective observational study was based on data transmitted routinely by the 51 maternities to the regional organization responsible for newborn screening, in 2016 and first half of 2017.

RESULTS: All the facilities implemented the UNHS protocol, with 47 out of 51 using the recommended techniques. 99.7% of the 115,435 newborns were screened (excluding 0.2% of parental refusals). A retest was required for 10.2% of the babies. Among babies who didn't pass retest, 7.7% were lost to follow-up. 2.2% of the newborns were referred to diagnostic centers. The rate of T3 was 31.3% of newborns who did not pass retest. 88.6% of the infants passed T3. In the perinatal network making extensive use of T3 (75.8% versus 14.9% elsewhere), 0.6% of the infants were referred to a diagnostic center, versus 2.9% in the rest of the region (2016, $p < 0.001$). For 2016, the outcomes at 6 months revealed an overall hearing loss rate of 1.7‰ (4.7‰ for neonatal care unit babies), and bilateral hearing loss in 1.2‰.

CONCLUSION: In Rhône-Alpes, the national and regional objectives for UNHS were exceeded, although limiting the number of infants lost to follow-up remains essential. Repeating an automated test around 2-4 weeks after birth improves the program by decreasing the false positives of the screening. It considerably limits the number of infants referred to specialist centers, without increasing the number of patients lost to follow-up.

Brennan-Jones CG, McMahen C, Van Dalen EC.

Cochrane corner: platinum-induced hearing loss after treatment for childhood cancer.

Int J Audiol. 2019. Apr;58(4):181-184. doi: 10.1080/14992027.2018.1539808. Epub 2018 Dec 13.

ABSTRACT: This Cochrane Corner features the review entitled "Platinum-induced hearing loss after treatment for childhood cancer" published in 2016. In their review, van As et al. identified 13 cohort studies including 2837 participants with a hearing test after treatment with a platinum-based therapy for different types of childhood cancers. All studies had problems related to quality of the evidence. The reported frequency of hearing loss varied between 1.7% and 90.1% for studies that included a definition of hearing loss; none of the studies

provided data on tinnitus. Only two studies evaluated possible risk factors. One study found a higher risk of hearing loss in people treated with the combination of cisplatin plus carboplatin compared to treatment with cisplatin only and for exposure to aminoglycosides. The other found that age at treatment (lower risk in older children) and single maximum cisplatin dose (higher risk with an increasing dose) were significant predictors for hearing loss, while gender was not. This systematic review shows that children treated with platinum analogues are at risk of developing hearing loss, but the exact prevalence and risk factors remain unclear.

Butcher E, Dezateux C, Knowles RL.
Risk factors for permanent childhood hearing impairment.
Arch Dis Child. 2018 Nov 28. pii: archdischild-2018-315866. doi: 10.1136/archdischild-2018-315866. [Epub ahead of print]

OBJECTIVE: While several perinatal risk factors for permanent childhood hearing impairment (PCHI) are known, association with gestational length remains unclear. We hypothesised that shorter gestational length predicts higher PCHI risk.
DESIGN: 19 504 participants from the UK Millennium Cohort Study (born 2000-2002, prior to newborn screening).
METHODS: Multivariable discrete-time survival analysis to examine associations between parent-reported PCHI by age 11 years and gestational length, plus other prespecified factors.
RESULTS: PCHI affected 2.1 per 1000 children (95% CI 1.5 to 3.0) by age 11; however, gestational length did not predict PCHI risk (HR, 95% CI 1.00, 0.98 to 1.03 per day increase). Risk was increased in those with neonatal illness, with or without admission to neonatal care (6.33, 2.27 to 17.63 and 2.62, 1.15 to 5.97, respectively), of Bangladeshi or Pakistani ethnicity (2.78, 1.06 to 7.31) or born to younger mothers (0.92, 0.87 to 0.97 per year).
CONCLUSION: Neonatal illness, rather than gestational length, predicts PCHI risk. Further research should explore associations with ethnicity.

Cedars E, Kriss H, Lazar AA, Chan C, Chan DK.
Use of otoacoustic emissions to improve outcomes and reduce disparities in a community preschool hearing screening program.
PLoS One. 2018 Dec 10;13(12):e0208050. doi: 10.1371/journal.pone.0208050. eCollection 2018.

INTRODUCTION: Hearing loss substantially impacts pediatric development, and early identification improves outcomes. While intervening before school-entry is critical to optimize learning, early-childhood hearing screening practices are highly variable. Conditioned play audiometry (CPA) is the gold standard for preschool hearing screening, but otoacoustic emission (OAE) testing provides objective data that may improve screening outcomes.
OBJECTIVES: To compare outcomes of a community-based low-income preschool hearing program before and after implementation of OAE in a single-visit, two-tiered paradigm. We hypothesized that this intervention would reduce referral rates and improve follow-up while maintaining stable rates of diagnosed sensorineural hearing loss.
METHODS: We performed a cohort study of 3257 children screened from July 2014-June 2016. Department of Public Health data were analyzed pre- and post-implementation of second-line OAE testing for children referred on CPA screening with targeted follow-up by DPH staff. Primary outcomes included referral rates, follow-up rates, and diagnosis of sensorineural hearing loss.
RESULTS: Demographics, pure-tone pass rates, and incidence of newly-diagnosed permanent hearing loss were similar across years. After intervention, overall pass rates increased from 92% to 95% ($P = 0.0014$), while only 0.7% remained unable to be tested ($P<0.0001$). 5% of children were unable to be tested by CPA screening but passed OAE testing, obviating further evaluation. Referral rate decreased from 8% to 5% ($P = 0.0014$), and follow-up improved from 36% to 91% ($P<0.0001$). Identification of pathology in children with follow-up increased from 19% to over 50%. Further, disparities in pass rates and ability to test seen in Year 1 were eliminated in Year 2.
CONCLUSION AND RELEVANCE: In a community setting, implementation of second-line OAE screening for CPA referrals reduced referral rates, increased identification of hearing loss, reduced outcome disparities, and improved follow-up rates. This study provides lessons in how to improve outcomes and reduce disparities in early-childhood hearing screening.

Cetin SY, Erel S, Bas Aslan U.
The effect of Tai Chi on balance and functional mobility in children with congenital sensorineural hearing loss.
Disabil Rehabil. 2019 Jan 9:1-8. doi: 10.1080/09638288.2018.1535629. [Epub ahead of print]

BACKGROUND: The aim of the study was to examine the effect of Tai Chi on balance and functional mobility in children with congenital sensorineural hearing loss.
METHODS: The study included 39 children, aged 10-14 years, with congenital sensorineural hearing loss. The

participants were divided into three groups as the Tai Chi group, conventional exercise group, and control group. The Tai Chi group and the conventional exercise group received a 1-h exercise program twice a week for 10 weeks. The balance function of the children was assessed using the Pediatric Balance Scale, the balance subtest of Bruininks-Oseretsky Test 2-Short Form, and the Functional Reach Test. The Timed Up and Go Test and the Timed Up and Down Stairs Test were used to assess functional mobility. The Wilcoxon rank, Kruskal-Wallis. and Mann-Whitney U-tests were used for statistical analyses.
RESULTS: When the pre-training values of the groups were compared, with the exception of the Timed Up and Go test, there was no statistically significant difference with respect to demographic data, balance, and functional mobility parameters ($p > 0.05$). After training, the overall balance and functional mobility tests improved compared to pre-training values in both the Tai Chi and conventional exercise groups ($p < 0.05$). When the post-training values were compared between the groups, with the exception of the Functional Reach Test and the Timed Up and Down Stairs Test, the results of both exercise groups were superior to those of the control group ($p < 0.05$).
CONCLUSIONS: The results of this study indicate that Tai Chi and conventional exercise programs have positive effects on balance and functional mobility in children with congenital sensorineural hearing loss. However, no superiority of Tai Chi or the conventional exercise programs was determined over the other. Both Tai Chi and conventional exercise programs could be used to improve balance and functional mobility in children with congenital sensorineural hearing loss. Implications for rehabilitation Tai Chi and conventional exercises are effective on balance in children with congenital sensorineural hearing loss. Tai Chi and conventional exercises are effective on functional mobility in children with congenital sensorineural hearing loss. Tai Chi may be added to the rehabilitation program for children with congenital sensorineural hearing loss.

Chan KH, Dreith S, Uhler KM, Tallo V, Lucero M, De Jesus J, Simões EA.
Large-scale otoscopic and audiometric population assessment: A pilot study.
Int J Pediatr Otorhinolaryngol. 2019 Feb;117:148-152. doi: 10.1016/j.ijporl.2018.11.033. Epub 2018 Nov 30.
OBJECTIVE: Large-scale otoscopic and audiometric assessment of populations is difficult due to logistic impracticalities, particularly in low- and middle-income countries (LMIC). We report a novel assessment methodology based on training local field workers, advances in audiometric testing equipment and cloud-based technology.
METHODS: Prospective observational study in Bohol, Philippines. A U.S. otolaryngologist/audiologist team trained 5 local nurses on all procedures in a didactic and hands-on process. An operating otoscope (Welch-Allyn®) was used to clear cerumen and view the tympanic membrane, images of which were recorded using a video otoscope (JedMed®). Subjects underwent tympanometry and distortion product otoacoustic emission (DPOAE) (Path Sentiero®), and underwent screening audiometry using noise cancelling headphones and a handheld Android device (HearScreen®). Sound-booth audiometry was reserved for failed subjects. Data were uploaded to a REDCap database. Teenage children previously enrolled in a 2000-2004 Phase 3 pneumococcal conjugate vaccine trial, were the subjects of the trainees.
RESULTS: During 4 days of training, 47 Filipino children (M/F = 28/19; mean/median age = 14.6/14.6 years) were the subjects of the trainee nurses. After the training, all nurses could perform all procedures independently. Otoscopic findings by ears included: normal (N = 77), otitis media with effusion (N = 2), myringosclerosis (N = 5), healed perforation (N = 6), perforation (N = 2) and retraction pocket/cholesteatoma (N = 2). Abnormal audiometric findings included: tympanogram (N = 4), DPOAE (N = 4) and screening audiometry (N = 0).
CONCLUSION: Training of local nurses has been shown to be robust and this methodology overcomes challenges of distant large-scale population otologic/audiometric assessment.

Chang YS, Ryu G, Kim K, Cho YS.
Normative wideband absorbance measures in healthy neonates in Korea: A preliminary study.
Int J Pediatr Otorhinolaryngol. 2019 Feb;117:6-11. doi: 10.1016/j.ijporl.2018.11.012. Epub 2018 Nov 11.
INTRODUCTION: The usefulness of wideband absorbance (WBA) in newborns is well-demonstrated. However, it is still not clear whether there might be a difference according to ethnicity with respect to ambient WBA; therefore, further investigation is necessary to evaluate ethnic-specific normative WBA values in newborns.
METHODS: Twenty-one newborns (41 ears) were recruited from the well-baby nursery at a tertiary referral center. All newborn infants who were born at 38 weeks' to 41 weeks' gestation with a normal birth weight (range: 2.5-4.5 kg) and who passed a newborn hearing screening test with distortion product otoacoustic emissions were enrolled. Ambient absorbance values were measured on frequencies ranging from 226 Hz to 6300 Hz (i.e., 250 Hz, 315 Hz, 400 Hz, 500 Hz, 620 Hz, 800 Hz, 1000 Hz, 1250 Hz, 1600 Hz, 2000 Hz, 2500 Hz, 3150 Hz, 4000 Hz, 5000 Hz, and 6300 Hz). The results of median absorbance were compared with the WBA values of Caucasian infants and Korean adults.
RESULTS: he gestational age of the study group was 38 weeks ± 6.67 days. In a gender comparison, absorbance of female neonate was significantly higher at 3150 Hz, 4000 Hz, and 5000 Hz than in male. Based on the

test frequencies, the medians of the Korean infant WBA values and Caucasian infants are significantly different from one another, except at 1600 Hz, 3150 Hz, and 4000 Hz. The results of a median absorbance comparison between Korean infant and adults WBA values showed that the medians of the two studies were significantly different except at 1250 Hz.

CONCLUSION: We analyzed the normative WBA values measured at ambient pressures in Korean newborns. The comparative analysis between the normative values of two different ethnic groups may infer a possible difference in the normative WBA values. The absorbance from Korean infant ears is substantially different from that from adult's ears. A large-scale study is required to establish normative WBA values to be used for the screening of outer and middle ear status in newborns.

Chen K, Jiang H, Zong L, Wu X.

Side-related differences in sudden sensorineural hearing loss in children.

Int J Pediatr Otorhinolaryngol. 2018 Nov;114:5-8. doi: 10.1016/j.ijporl.2018.08.022. Epub 2018 Aug 22.

OBJECTIVE: Most studies on sudden sensorineural hearing loss (SSNHL) do not differentiate the outcomes within varied affected ears in children. The present study was designed to determine the clinical differences between unilateral and bilateral SSNHL in children.

METHODS: The clinical data, from a total of 101 pediatric patients with SSNHL, was retrospectively analyzed from January 2003 to December 2016. The main outcome measures included basic characteristics, etiology, clinical symptoms and treatment courses.

RESULTS: When the bilateral group (n = 28) was compared to the unilateral group (n = 73), neither gender nor onset of SSNHL was significantly different ($p > 0.05$ each); However, bilateral SSNHL tended to occur in younger ages (8.1 ± 4.0 yrs), with higher percentages of suspected etiologies (50%) and proportion of profound deafness (55.4%, $p < 0.05$ each). The short-term recovery rate was superior in the unilateral cases over the bilateral cases (37.0% vs. 12.5%, $p < 0.05$). Milder initial hearing threshold, early onset of treatment (5.6 ± 4.8 days) with unilateral involvement and an older age (11.3 ± 3.0 yrs) in bilaterally affected cases were associated with a better prognosis in this cohort. In addition, the unilateral group showed comparable outcomes, when sub-analyzed by comparison to that in either left- (n = 42) or right-sided (n = 31) SSNHL.

CONCLUSION: Although bilateral and unilateral pediatric SSNHL could cause partial to complete cochlear lesion, they may be relevant to distinct backgrounds. Our data also provides valuable information about demographics and outcomes of SSNHL in children.

Clemens E, Brooks B, de Vries ACH, van Grotel M, van den Heuvel-Eibrink MM, Carleton B.

A comparison of the Muenster, SIOP Boston, Brock, Chang and CTCAEv4.03 ototoxicity grading scales applied to 3,799 audiograms of childhood cancer patients treated with platinum-based chemotherapy.

PLoS One. 2019 Feb 14;14(2):e0210646. doi: 10.1371/journal.pone.0210646. eCollection 2019.

ABSTRACT: Childhood cancer patients treated with platinums often develop hearing loss and the degree is classified according to different scales globally. Our objective was to compare concordance between five well-known ototoxicity scales used for childhood cancer patients. Audiometric test results (n = 654) were evaluated longitudinally and graded according Brock, Chang, International Society of Pediatric Oncology (SIOP) Boston, Muenster scales and the U.S. National Cancer Institute Common Technology Criteria for Adverse Events (CTCAE) version 4.03. Adverse effects of grade 2, 3 and 4 are considered to reflect a degree of hearing loss sufficient to interfere with day-to-day communication (> = Chang grade 2a; > = Muenster grade 2b). We term this "deleterious hearing loss". A total number of 3,799 audiograms were evaluated. The prevalence of deleterious hearing loss according to the last available audiogram of each patient was 59.3% (388/654) according to Muenster, 48.2% (315/653) according to SIOP, 40.5% (265/652) according to Brock, 40.3% (263/652) according to Chang, and 57.5% (300/522) according to CTCAEv4.03. Overall concordance between the scales ranged from $\kappa = 0.636$ (Muenster vs. Chang) to $\kappa = 0.975$ (Brock vs. Chang). Muenster detected hearing loss the earliest in time, followed by Chang, SIOP and Brock. Generally good concordance between the scales was observed but there is still diversity in definitions of functional outcomes, such as differences in distribution levels of severity of hearing loss, and additional intermediate scales taking into account losses <40 dB as well. Regardless of the scale used, hearing function decreases over time and therefore, close monitoring of hearing function at baseline and with each cycle of platinum therapy should be conducted.

Coleman A, Cervin A.

Probiotics in the treatment of otitis media. The past, the present and the future.

Int J Pediatr Otorhinolaryngol. 2019 Jan;116:135-140. doi: 10.1016/j.ijporl.2018.10.023. Epub 2018 Oct 19.

ABSTRACT: Otitis media (OM) is one of the most common infectious diseases in children and the leading cause for medical consultations and antibiotic prescription in this population. The burden of disease associated with OM is greater in developing nations and indigenous populations where the associated hearing loss contributes to poor education and employment outcomes. Current treatment and prevention is largely focused on

vaccination and antibiotics. However, rates of OM, particularly in indigenous populations, remain high. With growing concerns regarding antibiotic resistance and antibiotic-associated complications, an alternative, more effective treatment is required. Administration of probiotics, both locally and systemically have been investigated for their ability to treat and prevent OM in children. This review explores the theoretical bases of probiotics, successful application of probiotics in medicine, and their use in the treatment and prevention of OM. We conclude that local administration of niche-specific probiotic bacteria that demonstrates the ability to inhibit the growth of otopathogens in vitro shows promise in the prevention and treatment of OM and warrants further investigation.

De Schrijver L, Topsakal V, Wojciechowski M, Van de Heyning P, Boudewyns A.
Prevalence and etiology of sensorineural hearing loss in children with down syndrome: A cross-sectional study.
Int J Pediatr Otorhinolaryngol. 2019 Jan;116:168-172. doi: 10.1016/j.ijporl.2018.10.048. Epub 2018 Nov 3.

BACKGROUND: The prevalence and causes of sensorineural hearing loss (SNHL) in children with Down syndrome (DS) are poorly delineated.
OBJECTIVE: To describe the prevalence, severity, laterality and underlying etiology of SNHL in a cohort of children with DS.
METHODS: A cross-sectional study was performed among all children with DS followed at the multidisciplinary Downteam of the Antwerp University Hospital. Patients' characteristics, risk factors for hearing loss, audiometric data and results of an etiological work-up were collected.
RESULTS: Among 291 patients in follow-up, 138 patients (47.4%) presented with hearing loss. In the majority this was caused by middle ear effusion and only 13 patients (4.5%) had sensorineural hearing loss, 7 boys and 6 girls with a mean age of 14.4 ± 7.4 years. Hearing loss was bilateral in 8 cases. Hearing loss severity was graded as mild in 38.5%, moderate in 30.8% and profound in 30.8% of the patients. An etiological work-up was completed in 9 children. Four patients presented with single sided deafness due to cochlear nerve deficiency. One patient had a genetic cause and in 2 patients the hearing loss was attributed to excessive noise exposure. The etiology of hearing loss was unknown in 6 patients.
CONCLUSION: Sensorineural hearing loss is uncommon in children with DS with a prevalence of 4.5%. Etiological work-up may allow identifying a specific underlying cause. Cochlear nerve deficiency was found in 4 children with DS and single sided deafness.

Dedhia K, Graham E, Park A.
Hearing Loss and Failed Newborn Hearing Screen.
Clin Perinatol. 2018 Dec;45(4):629-643. doi: 10.1016/j.clp.2018.07.004. Epub 2018 Sep 24.

ABSTRACT: Hearing loss is the most common congenital defect. With early diagnosis and intervention, we are able to improve speech and language outcomes in this population. In this article, we discuss the implications of the newborn hearing screen, as well as diagnostic interventions, management, and intervention, and the increasing role of congenital cytomegalovirus screening.

Deng Y, Sang S, Wen J, Liu Y, Ling J, Chen H, Cai X, Mei L, Chen X, Li M, Li W, Li T, He C, Feng Y.
Reproductive guidance through prenatal diagnosis and genetic counseling for recessive hereditary hearing loss in high-risk families.
Int J Pediatr Otorhinolaryngol. 2018 Dec;115:114-119. doi: 10.1016/j.ijporl.2018.08.026. Epub 2018 Sep 12.

OBJECTIVE: To evaluate the accuracy and validity of our protocol for prenatal diagnosis and genetic counseling in high-risk families at a clinic.
METHODS: Fifteen unrelated families with recessive nonsyndromic hearing loss (NSHL) in their family history and a positive attitude towards prenatal diagnosis were recruited in the present study. According to genetic information for each family, Sanger sequencing, fluorescence polymerase chain reaction (PCR)-based congenital deafness gene detection kit and multiple PCR-based target gene capture and high-throughput sequencing were used. Genetic counseling was offered to all participating families by genetic counselors and otologists. Prenatal diagnosis was provided to families with detected pathogenic mutations and who were expected to participate in subsequent prenatal diagnosis.
RESULTS: In this study, confirmed pathogenic mutations were detected in eight families, who were defined as high-risk families. These families all participated in prenatal diagnosis with positive attitudes. One novel variant (c.1687dupA) in the SLC264 gene was detected in a family. Through genetic counseling, the recurrence probability of NSHL in fetuses was 25% in six families, 0% in one family, and 50% in one family. The results of fetal DNA detection showed that one fetal variant was wild type, three were heterozygous mutations in SLC26A4, and one was a compound heterozygous mutation in SLC26A4. Two variants were heterozygous mutations in GJB2, and one was a homozygous mutation in GJB2. According to the test results for fetal DNA, prenatal diagnosis found that six fetuses had normal hearing, whereas two fetuses suffered from NSHL. After birth, six infants predicted to have normal hearing passed a newborn hearing screening test and two infants predicted to

have NSHL were diagnosed with NSHL and received cochlear implants.
CONCLUSION: Our protocol for prenatal diagnosis and genetic counseling provides detailed information that can assist couples in high-risk families in preparing for infant arrival and future family planning. For the affected neonates, prenatal diagnosis and genetic counseling achieve an "early screening, early diagnosis, early intervention" strategy.

Dev AN, Lohith U, Pascal B, Dutt CS, Dutt SN.
A questionnaire-based analysis of parental perspectives on pediatric cochlear implant (CI) re/habilitation services: a pilot study from a developing CI service in India.
Cochlear Implants Int. 2018 Nov;19(6):338-349. doi: 10.1080/14670100.2018.1489937. Epub 2018 Jun 29.
OBJECTIVE: To study parental perspectives on re/habilitation services offered for pediatric cochlear implant (CI) users at a non-profit organization in India.
METHODOLOGY: A non-standardized questionnaire comprising 46 items was created to understand perspectives of parents of pediatric CI users. Questions were designed to examine re/habilitation services from the angles of service delivery, parental stress levels, reasons for delay in obtaining services, sources of emotional support, concerns, and fears during each stage starting from diagnosis of hearing loss to CI surgery, re/habilitation services and parents' views of their children post-CI. The questionnaire was posed to 30 parents and responses were recorded and coded.
RESULTS AND DISCUSSION: ualitative and quantitative analyses based on parents' responses identified several factors that significantly influenced parental perspectives during each stage. The major factors delaying the decision to go for CI included a fear of surgery, lack of funds for CI and the subsequent re/habilitation process, and limited knowledge. Key concerns were the child's academic performance and social acceptance. Familial support played an important role during each stage. A significant reduction in the parental stress levels was observed following CI surgery. Parents indicated that local support for therapy, financial assistance and better guidance at each stage would substantially help in lowering stress levels.
CONCLUSIONS: The parental perspectives analyzed in this study can be utilized towards improving the quality of service delivery in terms of parental satisfaction and outcomes post-CI. Efforts should be taken to improve parental awareness, funding options, and access to re/habilitation services and social networks connecting similar parents.

DiNino M, O'Brien G, Bierer SM, Jahn KN, Arenberg JG.
The Estimated Electrode-Neuron Interface in Cochlear Implant Listeners Is Different for Early-Implanted Children and Late-Implanted Adults.
J Assoc Res Otolaryngol. 2019 Mar 25. doi: 10.1007/s10162-019-00716-4. [Epub ahead of print]
ABSTRACT: Cochlear implant (CI) programming is similar for all CI users despite limited understanding of the electrode-neuron interface (ENI). The ENI refers to the ability of each CI electrode to effectively stimulate target auditory neurons and is influenced by electrode position, neural health, cochlear geometry, and bone and tissue growth in the cochlea. Hearing history likely affects these variables, suggesting that the efficacy of each channel of stimulation differs between children who were implanted at young ages and adults who lost hearing and received a CI later in life. This study examined whether ENI quality differed between early-implanted children and late-implanted adults. Auditory detection thresholds and most comfortable levels (MCLs) were obtained with monopolar and focused electrode configurations. Channel-to-channel variability and dynamic range were calculated for both types of stimulation. Electrical field imaging data were also acquired to estimate levels of intracochlear resistance. Children exhibited lower average auditory perception thresholds and MCLs compared with adults, particularly with focused stimulation. However, neither dynamic range nor channel-to-channel threshold variability differed between groups, suggesting that children's range of perceptible current was shifted downward. Children also demonstrated increased intracochlear resistance levels relative to the adult group, possibly reflecting greater ossification or tissue growth after CI surgery. These results illustrate physical and perceptual differences related to the ENI of early-implanted children compared with late-implanted adults. Evidence from this study demonstrates a need for further investigation of the ENI in CI users with varying hearing histories.

Dumont J, Abouzayd M, Le Louarn A, Pondaven S, Bakhos D, Lescanne E.
Total and partial ossiculoplasty in children: Audiological results and predictive factors.
Eur Ann Otorhinolaryngol Head Neck Dis. 2019 Mar 14. pii: S1879-7296(19)30037-7. doi: 10.1016/j.anorl.2019.02.012. [Epub ahead of print]
OBJECTIVE: To assess ossiculoplasty results in children and screen for predictive factors of efficacy.
PATIENTS AND METHODS: Seventy five children undergoing ossiculoplasty between 2001 and 2014 in a pediatric ENT department were included. The following data were collected and analyzed: demographic data, surgical indication, history of tympanoplasty, contralateral ear status (healthy, affected), preoperative hearing

thresholds, surgical technique, intraoperative findings, and ossicular chain status at eardrum opening. Audiological results were reported according to American Academy of Otolaryngology-Head and Neck Surgery guidelines.

RESULTS: Forty eight patients were included in the total ossicular reconstruction prosthesis (TORP) group. Mean age at surgery was 9.9years. Mean follow up was 2.7years. Mean air-bone gap (ABG) closure to within 20dB was achieved in 40% of cases at medium term (12 to 18 months after surgery). Air conduction (AC) threshold ≤30dB was achieved in 68% of cases. AC threshold improved by 14.6dB and 8.7dB at medium and long-term follow-up, respectively. A significant correlation was found between success rate and absence of history of tympanoplasty. The success rate was higher for primary than for revision procedures. Twenty seven children were included in the partial ossicular reconstruction prosthesis (PORP) group. Mean age was 9.5years, and mean follow-up 2.6years. Mean air-bone gap (ABG) closure to within 20dB was achieved in 75% of cases at medium term. AC threshold ≤30dB was achieved in 75% of cases AC threshold improved by 9.3dB and 5dB at medium and long-term follow-up, respectively. No predictive factors for success were found in the PORP group.

CONCLUSION: The present study suggested that total ossiculoplasty leads to better results when performed in first-line. It also confirmed that functional outcome is better in partial than total ossicular reconstruction prosthesis.

El-Dessouky HM, Aziz AA, Sheikhany AR, ElMeshmeshy LM.

Validation of the Egyptian Arabic Assessment of Auditory Skills development using children with Cochlear Implants.

Int J Pediatr Otorhinolaryngol. 2019 Apr 2;122:52-59. doi: 10.1016/j.ijporl.2019.03.033. [Epub ahead of print]

INTRODUCTION: Audition is the gateway to spoken language, and infants' early accomplishments in acquiring the sound structure of their native language lays a critical ground work for subsequent learning. The development of pre-lingual auditory perceptual skills for cochlear implanted children is crucial for initial development of oral language.

OBJECTIVE: The aims of the present study were to validate the Egyptian Arabic Assessment of Auditory Skills, and to track the development of auditory skills in Egyptian children fitted with CI during the first three years post implantation.

METHODS: The study included 90 Arabic Egyptian children attending the phoniatric unit, Kasr El Aini hospital. Their chronological age ranged from 36 to 72 months. The study lasted for 18 months from July 2015 to January 2017. The children were divided into six groups according to their cochlear age i.e., amount of implant experience. An Arabic assessment chart of auditory skills was tailored that included six auditory skills' domains; detection, identification, short term auditory memory, supra-segmental discrimination, segmental discrimination and linguistic auditory processing. This chart was then used to develop an assessment tool which was then applied to all the study participants. All children had bilateral Sensorineural Hearing Loss (SNHL) since birth. None of the participants had prior Cochlear Implant (CI), but all had tried conventional hearing aids. All participants were implanted unilateral, with CI devices. All met selection criteria applied in the Egyptian national insurance committee for cochlear implantation.

RESULTS: All auditory skills domains improved with cochlear age. There was significant improvement between 1-6 and 7-12 months in the scores of the Detection (DET) domain. There was significant difference between 1-6 and 7-12 months, 7-12 and 13-18 months, 19-24 and 25-30 months in the scores of the Identification (IDENT) domain. Regarding the Short Term Auditory Memory (STAM) domain scores and the Supra-segmental Discrimination (SSD) domain scores there was significant difference between all the groups. Regarding the Segmental Discrimination (SGD) domain scores, there was significant difference between group 1-6 and 7-12 months, 7-12 and 13-18 months, 19-24 and 25-30 months, 25-30 and 31-36 months. Regarding the Linguistic Auditory Processing (LAP) domain, there was significant difference between group 1-6 and 7-12 months, 7-12 and 13-18 months, 25-30 and 31-36 months.

CONCLUSIONS: Children fitted with Cochlear Implants (CIs) appeared to show improvement in acquisition of auditory skills over a period of three years that followed a hierarchy of development dependent on the cochlear age.

Faes J, Gillis S.

Auditory brainstem implantation in children with hearing loss: Effect on speech production.

Int J Pediatr Otorhinolaryngol. 2019 Apr;119:103-112. doi: 10.1016/j.ijporl.2019.01.014. Epub 2019 Jan 14.

ABSTRACT: Auditory brainstem implantation (ABI) is a recent technique in children's hearing restoration. Up till now the focus in the literature has mainly been the perceptual outcomes after implantation, whereas the effect of ABI on spoken language is still an almost unexplored area of research. This study presents a one-year follow-up of the volubility of two children with ABI. The volubility of signed and oral productions is investigated and oral productions are examined in more detail. Results show clear developmental trends in both children, indicating a beneficial effect of ABI on spoken language development.

Farrell AN, Landry AM, Yee ME, Leu RM, Goudy SL.
Sensorineural hearing loss in children with cell disease.
Int J Pediatr Otorhinolaryngol. 2019 Mar;118:110-114. doi: 10.1016/j.ijporl.2018.12.002. Epub 2018 Dec 5.

INTRODUCTION: Sensorineural hearing loss (SNHL) has been reported to occur at increased frequency in the pediatric sickle cell disease (SCD) population, likely secondary to ototoxic medication regimens and repeat sickling events that lead to end organ damage. Risk and protective factors of SNHL in this population are not fully characterized. The objective of this study was to describe audiology results in children with SCD and the prevalence and sequelae of SNHL.
METHODS: A comprehensive clinical database of 2600 pediatric SCD patients treated at 1 institution from 2010-16 was retrospectively reviewed to identify all patients who were referred for audiologic testing. Audiologic test results, patient characteristics, and SCD treatments were reviewed.
RESULTS: 181 SCD children (97 male, 153 HbSS) underwent audiologic testing, with 276 total audiology encounters, ranging 1-9 per patient. Mean age at first audiogram was 8.9 ± 5.2 years. 29.8% had prior cerebrovascular infarct and an additional 25.4% had prior abnormal transcranial Doppler screens documented at time of first audiogram. Overall, 13.3% had documented hearing loss, with 6.6% SNHL. Mean pure tone average (PTA) among patients with SNHL ranged from mild to profound hearing loss (Right: 43.3 ± 28.9, Left: 40.8 ± 29.7), sloping to more severe hearing loss at higher frequencies.
CONCLUSIONS: Hearing loss was identified in a significant subset of children with SCD and the hearing loss ranged from normal to profound. Though the overall prevalence of SNHL in SCD patients was low, baseline audiology screening should be considered.

Fitzgerald MP, Reynolds A, Garvey CM, Norman G, King MD, Hayes BC.
Hearing impairment and hypoxia ischaemic encephalopathy: Incidence and associated factors.
Eur J Paediatr Neurol. 2019 Jan;23(1):81-86. doi: 10.1016/j.ejpn.2018.10.002. Epub 2018 Oct 10.

OBJECTIVE: To establish the local incidence of hearing loss in newborns with Hypoxic Ischaemic Encephalopathy (HIE) and to identify associated risk factors.
STUDY DESIGN: Retrospective Cohort Study. Neonatal Intensive Care Unit (NICU) dual stage hearing screening protocol, including automated otoacoustic emissions (AOAE) and automated auditory brainstem response (AABR) testing.
RESULTS: 57 newborns received therapeutic hypothermia for HIE. Twelve babies (21%) died. Audiology data was incomplete in 3 babies. Complete data was available for 42 babies (male n = 24), 4 (9.5%) of whom had hearing impairment. The development of hearing loss was associated with abnormal blood glucose levels (p = 0.006), low Apgar score at 1 min (p = 0.0219) and evidence of multi organ dysfunction [high creatinine (p = 0.0172 and 0.0198) and raised liver transaminases (aspartate aminotransferase (AST) p = 0.0012, alanine aminotransferase (ALT) p = 0.0037)]. An association with gentamicin was not found.
CONCLUSION: This study confirms that hearing impairment is common in term infants who have undergone therapeutic hypothermia for moderate/severe HIE. Blood glucose should be monitored carefully in these infants and developmental surveillance should include formal audiology. Further larger studies are needed to clarify the role, if any, of hypothermia per se in causation of hearing loss and to fully identify risk factors for hearing impairment in this population.
WHAT IS NEW: The current study confirms that hearing impairment is common in term infants who have undergone therapeutic hypothermia for moderate/severe HIE. No association between gentamicin use and the development of hearing impairment was found however initial blood glucose outside the normal range was of significance. Other factors associated with hearing impairment were low Apgar scores, greater need for resuscitation and evidence of multi organ dysfunction (renal and liver failure).

Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Salamatmanesh M, Lee R, Fitzpatrick J.
Parent Report of Amplification Use in Children with Mild Bilateral or Unilateral Hearing Loss.
J Am Acad Audiol. 2019 Feb;30(2):93-102. doi: 10.3766/jaaa.17020. Epub 2018 Jan 15.

BACKGROUND: Amplification is considered to be one of the most important interventions for children with hearing loss. However, achieving consistent use of hearing technology in young children is an important problem, particularly when hearing loss is of mild degree. Little information is available about amplification use specifically for children with mild bilateral or unilateral hearing loss when such losses are targeted and identified early because of the availability of newborn hearing screening.
PURPOSE: We examined amplification use in a contemporary cohort of early-identified children with mild bilateral and unilateral hearing loss.
RESEARCH DESIGN: As part of the Mild and Unilateral Hearing Loss in Children Study, we collected parent reports on their child's use of amplification during the preschool years.
STUDY SAMPLE: A total of 69 children (38 unilateral and 31 bilateral mild) enrolled in the study from 2010 to 2015. Children entered the study at various ages between 12 and 36 mo of age and were followed up to age 48

mo. The median age of the children at enrollment was 16.5 mo (interquartile range [IQR] = 9.5, 26.8). Hearing loss was confirmed in these children at a median age of 3.6 mo (IQR = 2.4, 5.7).
DATA COLLECTION AND ANALYSIS: Baseline characteristics related to the child and family were collected through an intake form at study enrollment. Data on amplification fitting and use were collected via parent questionnaires at each assessment interval. Information from parent questionnaires was summarized descriptively and amplification use was grouped into categories. Through logistic regression, we examined the relationship between amplification use and laterality of hearing loss, sex, and maternal education.
RESULTS: Amplification was recommended for 59 (85.5%) children at a median age of 6.5 mo (IQR = 3.6, 21.2) and children were fitted at a median age of 10.9 mo (IQR = 6.0, 22.1). Based on parent report, hearing aid use was consistent for 39 (66.1%) of 59 children who had amplification recommended. Parent questionnaires showed very little change in use for most of the children over the study period. More children with bilateral hearing loss used their amplification consistently than those with unilateral hearing loss. After adjusting for maternal education and sex of the child, the odds for consistent use in children with mild bilateral loss was almost seven times higher (odds ratio = 6.75; 95% confidence interval = 1.84, 24.8) than for those with unilateral loss.
CONCLUSIONS: Although 85.5% of children with mild bilateral or unilateral hearing loss received amplification recommendations, only two-thirds achieved consistent use by age 3-4 yr based on parent report. Children with mild bilateral loss were more likely to use amplification during the preschool years than those with unilateral loss.

Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Nassrallah F.
Auditory and language outcomes in children with unilateral hearing loss.
Hear Res. 2019 Feb;372:42-51. doi: 10.1016/j.heares.2018.03.015. Epub 2018 Mar 13.
OBJECTIVES: Children with unilateral hearing loss (UHL) are being diagnosed at younger ages because of newborn hearing screening. Historically, they have been considered at risk for difficulties in listening and language development. Little information is available on contemporary cohorts of children identified in the early months of life. We examined auditory and language acquisition outcomes in a contemporary cohort of early-identified children with UHL and compared their outcomes at preschool age with peers with mild bilateral loss and with normal hearing.
DESIGN: As part of the Mild and Unilateral Hearing Loss in Children Study, we collected auditory and spoken language outcomes on children with unilateral, bilateral hearing loss and with normal hearing over a four-year period. This report provides a cross-sectional analysis of results at age 48 months. A total of 120 children (38 unilateral and 31 bilateral mild, 51 normal hearing) were enrolled in the study from 2010 to 2015. Children started the study at varying ages between 12 and 36 months of age and were followed until age 36-48 months. The median age of identification of hearing loss was 3.4 months (IQR: 2.0, 5.5) for unilateral and 3.6 months (IQR: 2.7, 5.9) for the mild bilateral group. Families completed an intake form at enrolment to provide baseline child and family-related characteristics. Data on amplification fitting and use were collected via parent questionnaires at each annual assessment interval. This study involved a range of auditory development and language measures. For this report, we focus on the end of follow-up results from two auditory development questionnaires and three standardized speech-language assessments. Assessments included in this report were completed at a median age of 47.8 months (IQR: 38.8, 48.5). Using ANOVA, we examined auditory and language outcomes in children with UHL and compared their scores to children with mild bilateral hearing loss and those with normal hearing.
RESULTS: On most measures, children with UHL performed poorer than those in the mild bilateral and normal hearing study groups. All children with hearing loss performed at lower levels compared to the normal hearing control group. However, mean standard scores for the normal hearing group in this study were above normative means for the language measures. In particular, children with UHL showed gaps compared to the normal hearing control group in functional auditory listening and in receptive and expressive language skills (three quarters of one standard deviation below) at age 48 months. Their performance in receptive vocabulary and speech production was not significantly different from that of their hearing peers.
CONCLUSIONS: Even when identified in the first months of life, children with UHL show a tendency to lag behind their normal hearing peers in functional auditory listening and in receptive and expressive language development.

Fu Y, Zha S, Lü N, Xu H, Zhang X, Shi W, Zha J.
Carrier frequencies of hearing loss variants in newborns of China: A meta-analysis.
J Evid Based Med. 2019 Feb;12(1):40-50. doi: 10.1111/jebm.12305. Epub 2018 Jul 2.
OBJECTIVE: The objective of this study was to review the carrier frequencies of hearing loss gene variants, such as GJB2, SLC26A4, and MT-RNR1 in newborns of China.
DESIGN: PubMed, Embase, BioCentral, CNKI, WanFang, and VIP databases were used for searching relevant literature studies published during the period of January 2007 and January 2016. Meta-analysis was performed

by using the R software. The estimated rate and its 95% confidence intervals (CI) of the relevant indexes in newborns were collected and calculated using a fixed-effects model or a random-effects model when appropriate.

RESULTS: In total, 35 of 958 published literature studies in Chinese and English were selected. The overall results showed that in newborns of China, the carrier frequencies of GJB2 variants (235 delC, 299 delAT) were 1.64% (95% CI 1.52% to 1.77%) and 0.33% (95% CI 0.19% to 0.51%); SLC26A4 variants (IVS7-2 A > G, 2168 A > G) were 1.02% (95% CI 0.91% to 1.15%) and 0.14% (95% CI 0.06% to 0.25%); MT-RNR1 variants (1555 A > G, 1449 C > T) were 0.20% (95% CI 0.17% to 0.23%) and 0.03% (95% CI 0.02% to 0.05%).

CONCLUSIONS: There are high carrier frequencies of GJB2 variants among newborns in China, followed by SLC26A4 and MT-RNR1 variants. In order to achieve “early detection, early diagnosis and early treatment” and reduce the incidence of hereditary hearing loss in offspring, a comprehensive combination of neonatal hearing screening and deafness gene detection should be recommended and implemented in China.

Funamura JL, Lee JW, McKinney S, Bayoumi AG, Senders CW, Tollefson TT.
Children with Cleft Palate: Predictors of Otologic Issues in the First 10 Years.
Otolaryngol Head Neck Surg. 2019 Jan 22:194599818825461. doi: 10.1177/0194599818825461. [Epub ahead of print]

OBJECTIVE: To evaluate the characteristics of children with cleft palate associated with persistent otologic issues in the first 10 years of life.

STUDY DESIGN: Case series with chart review.

SETTING: Single academic center.

SUBJECTS AND METHODS: Children born with cleft palate from 2003 to 2007 and treated by the UC Davis Cleft and Craniofacial Team between January 2003 and December 2017 were included in the study. Data from 143 patients were analyzed via Wilcoxon rank sum and Fisher exact tests for univariate analysis and logistic regression to determine adjusted odds ratios.

RESULTS: The median length of follow-up was 9.9 years, and the age at last ear examination was 10.7 years. At the last evaluation, unresolved otologic issues were common, with at least 1 ear having a tympanic membrane (TM) perforation (16.1%), a tympanostomy tube (36.2%), or conductive hearing loss (23.1%). After adjusting for demographic and clinical characteristics, history of palate revision or speech surgery was associated with having a TM perforation (P = .02). The only clinical variables associated with conductive hearing loss was the presence of a TM perforation (P < .01) or a genetic abnormality (P = .02). Severity of palatal clefting was not associated with specific otologic or audiologic outcomes after adjusting for other characteristics.

CONCLUSION: A large proportion of children with cleft palate have persistent otologic issues at age 10 years and would benefit from continued close monitoring well after the age when most children have normalized eustachian tube function. Prolonged otologic issues were not found to be associated with cleft type.

Goldsworthy RL, Markle KL.
Pediatric Hearing Loss and Speech Recognition in Quiet and in Different Types of Background Noise.
J Speech Lang Hear Res. 2019 Mar 25;62(3):758-767. doi: 10.1044/2018_JSLHR-H-17-0389.

Purpose: Speech recognition deteriorates with hearing loss, particularly in fluctuating background noise. This study examined how hearing loss affects speech recognition in different types of noise to clarify how characteristics of the noise interact with the benefits listeners receive when listening in fluctuating compared to steady-state noise.

Method: Speech reception thresholds were measured for a closed set of spondee words in children (ages 5-17 years) in quiet, speech-spectrum noise, 2-talker babble, and instrumental music. Twenty children with normal hearing and 43 children with hearing loss participated; children with hearing loss were subdivided into groups with cochlear implant (18 children) and hearing aid (25 children) groups. A cohort of adults with normal hearing was included for comparison.

Results: Hearing loss had a large effect on speech recognition for each condition, but the effect of hearing loss was largest in 2-talker babble and smallest in speech-spectrum noise. Children with normal hearing had better speech recognition in 2-talker babble than in speech-spectrum noise, whereas children with hearing loss had worse recognition in 2-talker babble than in speech-spectrum noise. Almost all subjects had better speech recognition in instrumental music compared to speech-spectrum noise, but with less of a difference observed for children with hearing loss.

Conclusions: Speech recognition is more sensitive to the effects of hearing loss when measured in fluctuating compared to steady-state noise. Speech recognition measured in fluctuating noise depends on an interaction of hearing loss with characteristics of the background noise; specifically, children with hearing loss were able to derive a substantial benefit for listening in fluctuating noise when measured in instrumental music compared to 2-talker babble.

Graham C, Seeley J, Gina A, Saman Y.
Mapping the content of mothers' knowledge, attitude and practice towards universal newborn hearing screening for development of a KAP survey tool.
PLoS One. 2019 Feb 20;14(2):e0210764. doi: 10.1371/journal.pone.0210764. eCollection 2019.

ABSTRACT: Understanding mother's knowledge, attitude and practice (KAP) of permanent childhood hearing impairment (PCHI) is essential for the success of universal newborn hearing screening (UNHS) as poor compliance and follow-up remains a global challenge. To determine content area for a questionnaire that measures PCHI-related KAP in rural mothers, we trained moderators who interviewed 145 pregnant women (17 groups) from 5 ante-natal clinics. Interviews were recorded, transcribed, summarised and analysed using thematic framework analysis. Four knowledge themes were identified: 1) PCHI was perceived as the malfunction of hearing leading to disability; 2) a poorly-responsive/communicative child may have PCHI; 3) lifestyle, hereditary and environmental factors are significant causes of PCHI; 4) medical management of PCHI was doubted, with some advocating birth and ancestral rituals. Two themes were identified for attitude: 1) beliefs that PCHI was emotionalised due to the negative lifelong impact on the child and family; 2) UNHS processes were favourable though some preferred other belief systems. Three themes were identified for practice: 1) doctors were the first choice followed by traditional healers; 2) willingness to continue follow-up although challenges exist; 3) minimal family support during consultation. The contextualised KAP of women regarding UNHS processes and PCHI provided content area for the design of a KAP tool.

Han JJ, Nguyen PD, Oh DY, Han JH, Kim AR, Kim MY, Park HR, Tran LH, Dung NH, Koo JW, Lee JH, Oh SH, Anh Vu H, Choi BY.
Elucidation of the unique mutation spectrum of severe hearing loss in a Vietnamese pediatric population.
Sci Rep. 2019 Feb 7;9(1):1604. doi: 10.1038/s41598-018-38245-4.

ABSTRACT: The mutational spectrum of deafness in Indochina Peninsula, including Vietnam, remains mostly undetermined. This significantly hampers the progress toward establishing an effective genetic screening method and early customized rehabilitation modalities for hearing loss. In this study, we evaluated the genetic profile of severe-to-profound hearing loss in a Vietnamese pediatric population using a hierarchical genetic analysis protocol that screened 11 known deafness-causing variants, followed by massively parallel sequencing targeting 129 deafness-associated genes. Eighty-seven children with isolated severe-to-profound non-syndromic hearing loss without family history were included. The overall molecular diagnostic yield was estimated to be 31.7%. The mutational spectrum for severe-to-profound non-syndromic hearing loss in our Vietnamese population was unique: The most prevalent variants resided in the MYO15A gene (7.2%), followed by GJB2 (6.9%), MYO7A (5.5%), SLC26A4 (4.6%), TMC1 (1.8%), ESPN (1.8%), POU3F4 (1.8%), MYH14 (1.8%), EYA1 (1.8%), and MR-RNR1 (1.1%). The unique spectrum of causative genes in the Vietnamese deaf population was similar to that in the southern Chinese deaf population. It is our hope that the mutation spectrum provided here could aid in establishing an efficient protocol for genetic analysis of severe-to-profound hearing loss and a customized screening kit for the Vietnamese population.

Hilditch C, Liersch B, Spurrier N, Callander EJ, Cooper C, Keir AK.
Does screening for congenital cytomegalovirus at birth improve longer term hearing outcomes?
Arch Dis Child. 2018 Oct;103(10):988-992. doi: 10.1136/archdischild-2017-314404. Epub 2018 Apr 28.

ABSTRACT: Currently, the diagnosis of congenital cytomegalovirus (cCMV) infection in most highly resourced countries is based on clinical suspicion alone. This means only a small proportion of cCMV infections are diagnosed. Identification, through either universal or targeted screening of asymptomatic newborns with cCMV, who would previously have gone undiagnosed, would allow for potential early treatment with antiviral therapy, ongoing audiological surveillance and early intervention if sensorineural hearing loss (SNHL) is identified. This paper systematically reviews published papers examining the potential benefits of targeted and universal screening for newborn infants with cCMV. We found that the treatment of these infants with antiviral therapy remains controversial, and clinical trials are currently underway to provide further answers. The potential benefit of earlier identification and intervention (eg, amplification and speech therapy) of children at risk of later-onset SNHL identified through universal screening is, however, clearer.

Hoffman HJ, Dobie RA, Losonczy KG, Themann CL, Flamme GA.
Kids Nowadays Hear Better Than We Did: Declining Prevalence of Hearing Loss in US Youth, 1966-2010.
Laryngoscope. 2018 Oct 5. doi: 10.1002/lary.27419. [Epub ahead of print]

OBJECTIVES/HYPOTHESIS: To investigate factors associated with hearing impairment (HI) in adolescent youths during the period 1966-2010.
STUDY DESIGN: Cross-sectional analyses of US sociodemographic, health, and audiometric data spanning 5 decades.
METHODS: Subjects were youths aged 12 to 17 years who participated in the National Health Examination

Survey (NHES Cycle 3, 1966-1970; n=6,768) and youths aged 12 to 19 years in the Third National Health and Nutrition Examination Survey (NHANES III, 1988-1994; n=3,057) and NHANES (2005-2010; n=4,374). HI prevalence was defined by pure-tone average (PTA)≥20 dB HL for speech frequencies (0.5, 1, 2, and 4 kHz) and high frequencies (3, 4, and 6 kHz). Multivariable logistic models were used to estimate the odds ratio (OR) and 95% confidence interval (CI).

RESULTS: Overall speech-frequency HI prevalence was 10.6% (95% CI: 9.7%-11.6%) in NHES, 3.9% (95% CI: 2.8%-5.5%) in NHANES III, and 4.5% (95% CI: 3.7%-5.4%) in NHANES 2005 to 2010. The corresponding high-frequency HI prevalences were 32.8% (95% CI: 30.8%-34.9%), 7.3% (95% CI: 5.9%-9.0%), and 7.9% (95% CI: 6.8%-9.2%). After adjusting for sociodemographic factors, overall high-frequency HI was increased twofold for males and cigarette smoking. Other significant risk factors in NHANES 2005 to 2010 included very low birth weight, history of ear infections/otitis media, ear tubes, fair/poor general health, and firearms use.

CONCLUSIONS: HI declined considerably between 1966 to 1970 and 1988 to 1994, with no additional decline between 1988 to 1994 and 2005 to 2010. Otitis media history was a significant HI risk factor each period, whereas very low birth weight emerged as an important risk factor after survival chances improved. Reductions in smoking, job-related noise, and firearms use may partially explain the reduction in high-frequency HI. Loud music exposure may have increased, but does not account for HI differences.

Hollanders JJ, Schaëfer N, van der Pal SM, Oosterlaan J, Rotteveel J, Finken MJJ; on behalf of the Dutch POPS-19 Collaborative Study Group.

Long-Term Neurodevelopmental and Functional Outcomes of Infants Born Very Preterm and/or with a Very Low Birth Weight.

Neonatology. 2019 Mar 5;115(4):310-319. doi: 10.1159/000495133. [Epub ahead of print]

BACKGROUND: Birth weight (BW) is often used as a proxy for gestational age (GA) in studies on preterm birth. Recent findings indicate that, in addition to perinatal outcomes, subjects born very preterm (VP; GA < 32 weeks) differ from those with a very low birth weight (VLBW; BW < 1,500 g) in postnatal growth up to their final height.

OBJECTIVE: To study whether neurodevelopmental and functional outcomes at the age of 19 years differ in VP and/or VLBW subjects.

METHODS: 705 19-year-old subjects from the Project on Preterm and Small-for-Gestational-Age Infants (POPS) cohort were classified as (1) VP+/VLBW+ (n = 354), (2) VP+/VLBW- (n = 144), or (3) VP-/VLBW+ (n = 207), and compared with regard to IQ as assessed with the Multicultural Capacity Test-intermediate level; neuromotor function using Touwen's examination of mild neurologic dysfunction; hearing loss; self- and parent-reported behavioral and emotional functioning; educational achievement and occupation; and self-assessed health using the Health Utilities Index and the London Handicap Scale.

RESULTS: VP+/VLBW- infants, on average, had 3.8-point higher IQ scores (95% confidence interval [CI] 0.5-7.1), a trend towards higher educational achievement, 3.3-dB better hearing (95% CI 1.2-5.4), and less anxious behavior, attention problems, and internalizing behavior than to VP+/VLBW+ subjects. VP-/VLBW+ infants reported 1.8 increased odds (95% CI 1.2-2.6) of poor health compared to VP+/VLBW+ subjects.

CONCLUSIONS: At the age of 19 years, subjects born VP+/VLBW+, VP+/VLBW-, and VP-/VLBW+ have different neurodevelopmental and functional outcomes, although effect sizes are small. Hence, the terms VP and VLBW are not interchangeable. We recommend, at least for industrialized countries, to base inclusion in future studies on preterm populations on GA instead of on BW.

Howell JB, Appelbaum EN, Armstrong MF, Chapman D, Dodson KM.

An Analysis of Risk Factors in Unilateral Versus Bilateral Hearing Loss.

Ear Nose Throat J. 2019 Apr 15:145561319840578. doi: 10.1177/0145561319840578. [Epub ahead of print]

ABSTRACT: A retrospective review of children with confirmed hearing loss identified through universal newborn hearing screening (UNHS) in Virginia from 2010 to 2014 was conducted in order to compare the incidence of Joint Committee on Infant Hearing (JCIH) risk factors in children with unilateral hearing loss (UHL) to bilateral hearing loss (BHL). Over the 5-year study period, 1004 children (0.20% of all births) developed a confirmed hearing loss, with 544 (51%) children having at least one JCIH risk factor. Overall, 18% of children with confirmed hearing loss initially passed UNHS. Of all children with risk factors, 226 (42%) demonstrated UHL and 318 (58%) had BHL. The most common risk factors for UHL were neonatal indicators (69%), craniofacial anomalies (30%), stigmata of HL syndromes (14%), and family history (14%). The most common risk factors in BHL were neonatal indicators (49%), family history (27%), stigmata of HL syndromes (19%), and craniofacial anomalies (16%). Children with the risk factor for positive family history were more likely to have BHL, while those with craniofacial anomalies were more likely to have UHL (P < .001). Neonatal indicators were the most commonly identified risk factor in both UHL and BHL populations. Children with UHL were significantly more likely to have craniofacial anomalies, while children with BHL were more likely to have a family history of hearing loss. Further studies assessing the etiology underlying the hearing loss and risk factor associations are warranted.

Iwanicka-Pronicka K, Ciara E, Piekutowska-Abramczuk D, Halat P, Pajdowska M, Pronicki M.
Congenital cochlear deafness in mitochondrial diseases related to RRM2B and SERAC1 gene defects. A study of the mitochondrial patients of the CMHI hospital in Warsaw, Poland.
Int J Pediatr Otorhinolaryngol. 2019 Mar 16;121:143-149. doi: 10.1016/j.ijporl.2019.03.015. [Epub ahead of print]

OBJECTIVES: Although hearing loss is a well-known symptom of mitochondria-related disorders, it is not clear how often it is a congenital and cochlear impairment. The Newborn Hearing Screening Program (NHSP) enables to distinguish congenital cochlear deafness from an acquired hearing deficit. The initial aim of the study was to research the frequency of the congenital cochlear hearing loss among patients with various gene defects resulting in mitochondrial disorders. The research process brought on an additional gain: basing on our preliminary study group of 80 patients, in 12 patients altogether we identified two defected genes responsible for mitochondrial disorders, whose carriers did not pass the NHSP. Finally, these patients were diagnosed with the congenital cochlear deafness.
MATERIAL AND METHODS: The results of the NHSP in the patients with mitochondrial disorders diagnosed in our tertiary reference center were analyzed. Only the cases with confirmed mutations were qualified for the study group. The NHSP database included 80 patients with mutations in 31 different genes: 25 nuclear-encoded and 6 mtDNA-encoded. We searched the literature for the presence of a congenital hearing impairment (CHI) in mitochondrial disorders caused by changes in 278 already known genes.
RESULTS: For 68 patients from the study group the NHSP test indicated a proper cochlear function and thus suggested normal hearing. For 12 mitochondrial patients, the NHSP test indicated the requirement for the further audiological diagnosis, and finally CHI was confirmed in 8 of them. This latter subset included patients with pathogenic variants in RRM2B and SERAC1, known as “deafness-causing genes”. Contrary to our initial expectations, the patients carrying mutations in other “deafness-causing genes”: MPV17, POLG, COX10, as well as other mitochondria-related genes, all reported in literature, did not indicate any CHI following the NHSP test.
CONCLUSION: Our study indicates that the cochlear CHI is a phenotypic feature of the RRM2B and SERAC1 related defects. The diagnosis of the CHI following the NHSP allows to early distinguish those defects from other mitochondria-related disorders in which the NHSP test result is correct. Wider studies are needed to assess the significance of this observation.

Jabbour N, Weinreich HM, Owusu J, Lehn M, Yueh B, Levine S.
Hazardous noise exposure from noisy toys may increase after purchase and removal from packaging: A call for advocacy.
Int J Pediatr Otorhinolaryngol. 2019 Jan;116:84-87. doi: 10.1016/j.ijporl.2018.10.028. Epub 2018 Oct 22.

OBJECTIVE: Previous studies identified hazardous noise levels from packaged toys. Sound levels may increase when packaging is removed and therefore, complicate the ability to accurately assess noise levels before purchase. The goal of this study was to evaluate how packaging affects the decibel (dB) level of toys by: 1) Assessing dB level of toys with and without packaging. 2) Evaluating the percentage of packaged and unpackaged toys that exceed a safety limit of 85 dB.
METHODS: Thirty-five toys were selected from the 2009-2011 Sight and Hearing Association (SHA) based on availability for purchase. Toys’ speakers were categorized as Exposed, Partially Exposed, or Covered, based on its packaging. The dB level of each toy was tested at 0 cm and 25 cm from the speaker using a handheld digital sound meter in a standard audiometric booth. T tests and ANOVA were performed to assess mean change in sound level before and after packaging removal.
RESULTS: Significant dB increases were noted after packaging was removed (mean change 11.9 dB at 0 cm; and 2.5 dB at 25 cm, $p < 0.001$). Sixty-four percentage of Covered toys ($n = 14$) had dB greater than 85 dB when packaged and this increased to 100% when unpackaged.
CONCLUSION: Many manufactured toys have hazardous sound levels. Caregivers and healthcare providers should be aware that toys tested in the store may actually be louder when brought home and removed from their packaging. Limits on and disclosure of dB level of toys should be considered nationally.

Jackson W, Taylor G, Selewski D, Smith PB, Tolleson-Rinehart S, Laughon MM.
Association between furosemide in premature infants and sensorineural hearing loss and nephrocalcinosis: a systematic review.
Matern Health Neonatol Perinatol. 2018 Nov 19;4:23. doi: 10.1186/s40748-018-0092-2. eCollection 2018.

ABSTRACT: Furosemide is a potent loop diuretic commonly and variably used by neonatologists to improve oxygenation and lung compliance in premature infants. There are several safety concerns with use of furosemide in premature infants, specifically the risk of sensorineural hearing loss (SNHL), and nephrocalcinosis/nephrolithiasis (NC/NL). We conducted a systematic review of all trials and observational studies examining the association between these outcomes with exposure to furosemide in premature infants. We searched MEDLINE, EMBASE, CINAHL, and clinicaltrials.gov. We included studies reporting either SNHL or NC/NL in premature

infants (<37 weeks completed gestational age) who received at least one dose of enteral or intravenous furosemide. Thirty-two studies met full inclusion criteria for the review, including 12 studies examining SNHL and 20 studies examining NC/NL. Only one randomized controlled trial was identified in this review. We found no evidence that furosemide exposure increases the risk of SNHL or NC/NL in premature infants, with varying quality of studies and found the strength of evidence for both outcomes to be low. The most common limitation in these studies was the lack of control for confounding factors. The evidence for the risk of SNHL and NC/NL in premature infants exposed to furosemide is low. Further randomized controlled trials of furosemide in premature infants are urgently needed to adequately assess the risk of SNHL and NC/NL, provide evidence for improved FDA labeling, and promote safer prescribing practices.

Judge PD, Jorgensen E, Lopez-Vazquez M, Roush P, Page TA, Moeller MP, Tomblin JB, Holte L, Buchman C.
Medical Referral Patterns and Etiologies for Children With Mild-to-Severe Hearing Loss.
Ear Hear. 2018 Dec 6. doi: 10.1097/AUD.0000000000000682. [Epub ahead of print]

OBJECTIVES: To (1) identify the etiologies and risk factors of the patient cohort and determine the degree to which they reflected the incidence for children with hearing loss and (2) quantify practice management patterns in three catchment areas of the United States with available centers of excellence in pediatric hearing loss.
DESIGN: Medical information for 307 children with bilateral, mild-to-severe hearing loss was examined retrospectively. Children were participants in the Outcomes of Children with Hearing Loss (OCHL) study, a 5-year longitudinal study that recruited subjects at three different sites. Children aged 6 months to 7 years at time of OCHL enrollment were participants in this study. Children with cochlear implants, children with severe or profound hearing loss, and children with significant cognitive or motor delays were excluded from the OCHL study and, by extension, from this analysis. Medical information was gathered using medical records and participant intake forms, the latter reflecting a caregiver's report. A comparison group included 134 children with normal hearing. A Chi-square test on two-way tables was used to assess for differences in referral patterns by site for the children who are hard of hearing (CHH). Linear regression was performed on gestational age and birth weight as continuous variables. Risk factors were assessed using t tests. The alpha value was set at $p < 0.05$.
RESULTS: Neonatal intensive care unit stay, mechanical ventilation, oxygen requirement, aminoglycoside exposure, and family history were correlated with hearing loss. For this study cohort, congenital cytomegalovirus, strep positivity, bacterial meningitis, extracorporeal membrane oxygenation, and loop diuretic exposure were not associated with hearing loss. Less than 50% of children underwent imaging, although 34.2% of those scanned had abnormalities identified. No single imaging modality was preferred. Differences in referral rates were apparent for neurology, radiology, genetics, and ophthalmology.
CONCLUSIONS: The OCHL cohort reflects known etiologies of CHH. Despite available guidelines, centers of excellence, and high-yield rates for imaging, the medical workup for children with hearing loss remains inconsistently implemented and widely variable. There remains limited awareness as to what constitutes appropriate medical assessment for CHH.

Kanabur P, Hubbard C, Jeyakumar A.
Clinical Guidelines in Pediatric Hearing Loss: Systemic Review Using the Appraisal of Guidelines for Research and Evaluation II Instrument.
Laryngoscope. 2018 Dec 8. doi: 10.1002/lary.27722. [Epub ahead of print]

OBJECTIVES: Despite the importance, impact, and prevalence of pediatric hearing loss (HL), there are very few published clinical practice guidelines (CPG) supporting the evaluation and management of pediatric patients with HL. Our objective was to appraise existing CPGs to ensure safe and effective practices.
METHODS: A literature search was conducted in PubMed, Google Scholar, EBSCO, as well as a manual Google search. Three independent assessors using the Appraisal of Guidelines for Research and Evaluation II (AGREE II) instrument evaluated CPGs related to HL in children. Standardized domain scores were calculated for each guideline.
RESULTS: A total of four guidelines met the inclusion criteria and were appraised. Scope and purpose achieved a high median score of 83%. Stakeholder involvement, clarity of presentation, and editorial independence achieved intermediate scores of 67%, 54%, and 50%, respectively. The areas that required most improvement and achieved low scores were rigor of development and applicability, with scores of 22% and 38%, respectively. Based on the AGREE II measures, the four guidelines had domain scores less than 60% for each domain, and without modification no guideline could be recommended.
CONCLUSIONS: Based on the AGREE II, the qualities of CPGs for pediatric HL have several shortcomings, and the need for a comprehensive CPG remains. Rigor of development and applicability present the greatest opportunities for improvement of these CPGs.

Kanji A, Krabbenhoft K.
Audiological follow-up in a risk-based newborn hearing screening programme: An exploratory study of the influencing factors.
S Afr J Commun Disord. 2018 Oct 25;65(1):e1-e7. doi: 10.4102/sajcd.v65i1.587.

BACKGROUND: Follow-up return rate in Early Hearing Detection and Intervention (EHDI) programmes is of specific importance as it ensures that benchmarks are met and that no child with suspected hearing loss is left unidentified.
OBJECTIVES: The aim of this study was to determine the factors influencing audiological follow-up of high-risk infants in a risk-based newborn hearing screening programme.
METHOD: A non-experimental, exploratory, qualitative research design was employed. Purposive sampling was used. The study was conducted at a secondary level hospital in the public health care sector in South Africa. Participants comprised 10 caregivers (age range 26-40 years) of infants who had been enrolled in a risk-based newborn hearing screening programme, and returned for follow-up appointments. Data were collected using semi-structured interviews. Responses were recorded by the researcher and a colleague to ensure rigour and trustworthiness of findings. Data were analysed using thematic analysis for open-ended questions and descriptive statistics for the closed-ended questions.
RESULTS: The most common positive contributors that facilitated participants› attendance at follow-up appointments were: having friendly audiologists; a clear line of communication between caregiver and audiologist and a reminder of the appointment. The most significant perceived challenge that participants described in returning for the follow-up appointment was living in far proximity from the hospital.
CONCLUSION: Findings of the study revealed that influencing factors on follow-up return rate are demographic, socio-economic, and interpersonal in nature and further suggested the need for an all-inclusive appointment day. It may be of importance to not only look at what is being done to improve the follow-up return rate but also how it should be done in terms of professional-to-patient communication and interactions.

Kanji A, Khoza-Shangase K, Moroe N.
Newborn hearing screening protocols and their outcomes: A systematic review.
Int J Pediatr Otorhinolaryngol. 2018 Dec;115:104-109. doi: 10.1016/j.ijporl.2018.09.026. Epub 2018 Sep 25.

OBJECTIVE: To conduct a review of the most current research in objective measures used within newborn hearing screening protocols with the aim of exploring the actual protocols in terms of the types of measures used and their frequency of use within a protocol, as well as their outcomes in terms of sensitivity, specificity, false positives, and false negatives in different countries worldwide.
METHODS: A systematic literature review was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analysis. Electronic databases such as PubMed, Google Scholar and Science Direct were used for the literature search. A total of 422 articles were identified, of which only 15 formed part of the current study. The 15 articles that met the study's criteria were reviewed. Pertinent data and findings from the review were tabulated and qualitatively analysed under the following headings: country; objective screening and/or diagnostic measures; details of screening protocol; results (including false positive and negative findings, sensitivity and/or specificity), conclusion and/or recommendations. These tabulated findings were then discussed with conclusions and recommendations offered.
RESULTS: Findings reported in this paper are based on a qualitative rather than a quantitative analysis of the reviewed data. Generally, findings in this review revealed firstly, that there is a lack of uniformity in protocols adopted within newborn hearing screening. Secondly, many of the screening protocols reviewed consist of two or more tiers or stages, with transient evoked otoacoustic emissions (TEOAEs) and automated auditory brainstem response (AABR) being most commonly used. Thirdly, DPOAEs appear to be less commonly used when compared to TEOAEs. Lastly, a question around routine inclusion of AABR as part of the NHS protocol remains inconclusively answered.
CONCLUSIONS: There is sufficient evidence to suggest that the inclusion of AABR within a NHS programme is effective in achieving better hearing screening outcomes. The use of AABR in combination with OAEs within a test-battery approach or cross-check principle to screening is appropriate, but the inclusion of AABR to facilitate appropriate referral for diagnostic assessment needs to be systematically studied.

Kapitanova M, Knebel JF, El Ezzi O, Artaz M, de Buys Roessingh AS, Richard C.
Influence of infancy care strategy on hearing in children and adolescents: A longitudinal study of children with unilateral lip and /or cleft palate.
Int J Pediatr Otorhinolaryngol. 2018 Nov;114:80-86. doi: 10.1016/j.ijporl.2018.08.031. Epub 2018 Aug 27.

OBJECTIVES: To evaluate the relation between ventilation tube insertion, otitis media with effusion duration and otologic outcomes in unilateral cleft lip and/or cleft palate children from infancy to teenage age.
DESIGN AND POPULATION: Retrospective longitudinal charts review of patients from the multidisciplinary cleft team of the University Hospital of Lausanne over a 30-year period. 146 charts from consecutive patients with

non-syndromic unilateral cleft lip and/or cleft palate who were born between January 1986 and January 2003 were included.
RESULTS: The earlier in life a cleft child experience his first otitis media with effusion (OME), the worse his long-term hearing will be. Along with the age of onset of OME, we disclosed an influence of the duration of OME without ventilation tube (VT) insertion on short and long-term hearing outcomes. Different patterns were observed between cleft palate (CP) and cleft lip palate children (CLP), with a higher incidence of otitis media with effusion for the CLP group than the CP group. Direct positive relationship between VT insertion and hearing were disclosed and evaluation of long-term complications did not reveal significant relation with VT insertion. Of note, OME in CLP children led to a higher rate (but not statistically significant) of chronic ear complications than in the CP group, that may indicate more persistent OME or different adverse effect on the middle ear mucosa between CP and CLP children.
CONCLUSIONS: Individualized counseling should take into account different factors such as the type of cleft, the age of onset of OME and duration of OME, keeping in mind the adverse effect of persistent middle ear fluid. In the present report, results prone an early ventilation tube insertion to prevent short and long-term injury to the middle ear homeostasis, hearing loss and related issues.

Karanth TK, Whittemore KR.
Middle-ear disease in children with cleft palate.
Auris Nasus Larynx. 2018 Dec;45(6):1143-1151. doi: 10.1016/j.anl.2018.04.012.
OBJECTIVE: The objective of this review is to summarize all aspects of middle ear diseases in children with cleft palate (CP).
METHODS: PubMed, Scopus, The Cumulative Index to Nursing and Allied Health Literature (CINAHL) and The Cochrane Library were searched for English-language randomized control trials (RCTs), meta-analyses, systematic reviews and observational studies published through 31st July 2017.
RESULTS: Epidemiology and pathogenesis of middle ear diseases in children with cleft palate have been discussed in this review. Methods of Evaluation, CP surgeries, complications and follow up have been detailed for the same.
CONCLUSION: Evaluation of middle-ear disease in children with CP begins at birth by a newborn hearing screen. Tympanometry and otoscopy helps screen for middle-ear disease during follow-up visits. Ventilation tube may be placed when indicated based on the patient's clinical course and presentation. Long-term follow up should be provided to look for the development of cholesteatoma.

Khoza-Shangase K.
Early hearing detection and intervention in South Africa: Exploring factors compromising service delivery as expressed by caregivers.
Int J Pediatr Otorhinolaryngol. 2019 Mar;118:73-78. doi: 10.1016/j.ijporl.2018.12.021. Epub 2018 Dec 18.
AIM: The main aim of this study was to explore factors compromising early intervention (EI) service delivery to hearing impaired children in South Africa, as expressed by their caregivers.
METHODS: Within a qualitative survey design, a sample of 19 hearing impaired children's caregivers completed structured self-administered questionnaires on factors that they perceive compromise EI for their children. These caregivers included mothers, fathers, grandparents, and legal guardians or adoptive parents of children with hearing impairment. Descriptive analysis of the data was undertaken.
RESULTS: Findings indicated various factors compromising EI as reported by caregivers. These included limited availability of appropriate schools and health care facilities for their hearing impaired children; long distances between the few services that are available and the places of residence of the service users; significant costs linked to the services (such as medical expenses, boarding school facilities costs); limited skills and knowledge of professionals and teachers regarding hearing impairment; inconsistent and conflicting professional opinions about the child's diagnosis and treatment; as well as limited community awareness about hearing impairment along with services available for hearing impaired children.
CONCLUSION: These findings have important clinical, training, policy, and advocacy implications within the South African context; if both access to and success within the EI services will be successful.

Kim SY, Choi BY, Jung EY, Park H, Yoo HN, Park KH.
Risk factors for failure in the newborn hearing screen test in very preterm twins.
Pediatr Neonatol. 2018 Dec;59(6):586-594. doi: 10.1016/j.pedneo.2018.01.014. Epub 2018 Jan 31.
BACKGROUND: We aimed to identify prenatal and postnatal risk factors associated with abnormal newborn hearing screen (NHS) results and subsequently confirmed sensorineural hearing loss (SNHL) in preterm twin neonates.
METHODS: Electronic medical records of 159 twin neonates who were born alive after ≤32 weeks were retrospectively reviewed for hearing loss in both ears. Histopathologic examination of the placenta was

performed and clinical data, including method of conception and factors specific to twins, were retrieved from a computerized perinatal database. The main outcome measure was failure to pass the NHS test. The generalized estimation equations model was used for twins.

RESULTS: Thirty-two neonates (20.1%) had a "refer" result, and, on the confirmation test, permanent SNHL was identified in 4.4% (7/159) of all neonates. Neonates who had a "refer" result on the NHS test were more likely to be of lower birth weight, more likely to have been conceived with the use of in vitro fertilization (IVF), and more likely to have higher rates of intraventricular hemorrhage (IVH) and bronchopulmonary dysplasia. However, monochorionic placentation, death of the co-twin, or being born first was not associated with a "refer" result on the NHS test. Multivariable logistic regression revealed that conception after IVF and the presence of IVH were the only variables to be statistically significantly associated with "refer" on the NHS test. No parameters studied were found to be significantly different between the SNHL and no SNHL groups, probably because of the relatively small number of cases of SNHL.

CONCLUSION: In preterm twin newborns, IVF and the presence of IVH were independently associated with an increased risk of abnormal NHS results, whereas the factors specific to twins were not associated with abnormal NHS results.

Kitao K, Mutai H, Namba K, Morimoto N, Nakano A, Arimoto Y, Sugiuchi T, Masuda S, Okamoto Y, Morita N, Sakamoto H, Shintani T, Fukuda S, Kaga K, Matsunaga T.

Deterioration in Distortion Product Otoacoustic Emissions in Auditory Neuropathy Patients With Distinct Clinical and Genetic Backgrounds.

Ear Hear. 2019 Jan/Feb;40(1):184-191. doi: 10.1097/AUD.0000000000000586.

OBJECTIVES: Auditory neuropathy (AN) is a clinical disorder characterized by the absence of auditory brainstem response and presence of otoacoustic emissions. A gradual loss of otoacoustic emissions has been reported for some cases of AN. Such cases could be diagnosed as cochlear hearing loss and lead to misunderstanding of the pathology when patients first visit clinics after the loss of otoacoustic emissions. The purpose of this study was to investigate the time course of changes in distortion product otoacoustic emissions (DPOAEs) in association with patients' genetic and clinical backgrounds, including the use of hearing aids.

DESIGN: DPOAE measurements from 31 patients with AN were assessed. Genetic analyses for GJB2, OTOF, and mitochondrial m.1555A> G and m.3243A> G mutations were conducted for all cases, and the analyses for CDH23 and OPA1 were conducted for the selected cases. Patients who were younger than 10 years of age at the time of AN diagnosis were designated as the pediatric AN group (22 cases), and those who were 18 years of age or older were designated as the adult AN group (9 cases). DPOAE was measured at least twice in all patients. The response rate for DPOAEs was defined and analyzed.

RESULTS: The pediatric AN group comprised 10 patients with OTOF mutations, 1 with GJB2 mutations, 1 with OPA1 mutation, and 10 with indefinite causes. Twelve ears (27%) showed no change in DPOAE, 20 ears (46%) showed a decrease in DPOAE, and 12 ears (27%) lost DPOAE. Loss of DPOAE occurred in one ear (2%) at 0 years of age and four ears (9%) at 1 year of age. The time courses of DPOAEs in patients with OTOF mutations were divided into those with early loss and those with no change, indicating that the mechanism for deterioration of DPOAEs includes not only the OTOF mutations but also other common modifier factors. Most, but not all, AN patients who used hearing aids showed deterioration of DPOAEs after the start of using hearing aids. A few AN patients also showed deterioration of DPOAEs before using hearing aids. The adult AN group comprised 2 patients with OPA1 mutations, 2 with OTOF mutations, and 5 with indefinite causes. Four ears (22%) showed no change in DPOAE, 13 ears (72%) showed a decrease, and one ear (6%) showed a loss of DPOAE. Although the ratio of DPOAE decrease was higher in the adult AN group than in the pediatric AN group, the ratio of DPOAE loss was lower in the adult AN group. DPOAE was not lost in all four ears with OPA1 mutations and in all four ears with OTOF mutations in the adult group.

CONCLUSIONS: DPOAE was decreased or lost in approximately 70% of pediatric and about 80% of adult AN patients. Eleven percent of pediatric AN patients lost DPOAEs by 1 year of age. Genetic factors were thought to have influenced the time course of DPOAEs in the pediatric AN group. In most adult AN patients, DPOAE was rarely lost regardless of the genetic cause.

Komori K, Komori M, Eitoku M, Joelle Muchanga SM, Ninomiya H, Kobayashi T, Suganuma N; Japan Environment and Children's Study (JECS) Group.

Verbal abuse during pregnancy increases frequency of newborn hearing screening referral: The Japan Environment and Children's Study.

Child Abuse Negl. 2019 Apr;90:193-201. doi: 10.1016/j.chiabu.2019.01.025. Epub 2019 Feb 23.

BACKGROUND: Verbal abuse during pregnancy has a greater impact than physical and sexual violence on the incidence of postnatal depression and maternal abuse behavior towards their children. In addition, exposure of children (aged 12 months to adolescence) to verbal abuse from their parents exerts an adverse impact to the children's auditory function. However, the effect of verbal abuse during pregnancy on fetal auditory function has

not yet been thoroughly investigated.
OBJECTIVE: The objective of the study was to examine the relationship between intimate partner verbal abuse during pregnancy and newborn hearing screening (NHS) referral, which indicates immature or impaired auditory function.
PARTICIPANTS AND SETTING: The Japan Environment and Children's Study is an ongoing nationwide population-based birth-cohort study designed to determine environmental factors during and after pregnancy that affect the development, health, or wellbeing of children. Pregnant women living in 15 areas of Japan were recruited between January 2011 and March 2014.
METHODS: Multiple imputation for missing data was performed, followed by multiple logistic regression using 16 confounding variables.
RESULTS: Of 104,102 records in the dataset, 79,985 mother-infant pairs submitted complete data for questions related to verbal and physical abuse and the results of NHS. Of 79,985 pregnant women, 10,786 (13.5%) experienced verbal abuse and 978 (1.2%) experienced physical abuse. Of 79,985 newborns, 787 (0.98%) received a NHS referral. Verbal abuse was significantly associated with NHS referral (adjusted odds ratio: 1.44; 95% confidence interval: 1.05-1.98).
CONCLUSIONS: Verbal abuse should be avoided during pregnancy to preserve the newborn's auditory function.

Lago MRR, Fernandes LDC, Lyra IM, Ramos RT, Teixeira R, Salles C, Ladeia AMT.
Sensorineural hearing loss in children with sickle cell anemia and its association with endothelial dysfunction.
Hematology. 2018 Dec;23(10):849-855. doi: 10.1080/10245332.2018.1478494. Epub 2018 May 28.
OBJECTIVES: To investigate the prevalence of sensorineural hearing loss (SNHL) in children and adolescents with sickle cell anemia (SCA) and its association with endothelial dysfunction (ED).
METHODS: Fifty-two participants with stable SCA and 44 apparently healthy (AA genotype) participants aged 6-18 years were evaluated for pure tone audiometry and endothelial function using ultrasonographic imaging of the brachial artery to assess flow-mediated dilation (FMD). Laboratory analysis of the lipid profile and C-reactive protein levels was performed.
RESULTS: In the SCA group, 15 (28.8%) patients presented with SNHL. The FMD values were reduced in the SCA with SNHL group compared with the SCA without SNHL and healthy groups. Logistic regression analysis showed that FMD was associated with SNHL independent of the lipid profile and SCA characteristics (odds ratio [95% confidence interval] = 0.614 [0.440-0.858]; p = 0.004).
DISCUSSION: SNHL is a common complication in SCA; furthermore, this study identified a significant association between ED and SNHL. Damage to the vascular endothelium because of inflammation in SCA reduced blood flow in the inner ear. Thus, this circulatory disorder culminates in vaso-occlusive process and induces auditory disorders, such as SNHL.

Lee ER, Chan DK.
Implications of dried blood spot testing for congenital CMV on management of children with hearing loss: A preliminary report.
Int J Pediatr Otorhinolaryngol. 2019 Apr;119:10-14. doi: 10.1016/j.ijporl.2018.12.029. Epub 2018 Dec 21.
INTRODUCTION: Non-genetic, congenital sensorineural hearing loss (cSNHL) is commonly caused by congenital CMV infection (cCMV). Hearing loss related to cCMV is variable in degree, often progressive, and can affect one or both ears.
OBJECTIVES: We sought to examine the outcomes of DBS testing in California, and the hearing outcomes of cCMV-positive children.
METHODS: This is a retrospective study of patients with SNHL of unknown etiology aged 6 months to 17 years old presenting to a tertiary care pediatric center and evaluated for cCMV by DBS testing.
RESULTS: 14 children (228 ears) with SNHL of unknown origin were included. 6/114 (5.3%) tested positive for cCMV versus 108/114 (94.7%), who tested negative. None of the cCMV-positive children had symmetric bilateral hearing loss, compared with 56.5% (61/108) of cCMV-negative children ($p < 0.05$). cCMV-positive children were more likely to have profound SNHL in the worse-hearing ear (5/6 (83%) vs 16/108 (14.9%) of cCMV-negative children, $p < 0.001$). 86% (5/6) exhibited progressive hearing loss, including progression or new-onset hearing loss in the previously better hearing ear. 3 of the 6 children with cCMV underwent CI.
CONCLUSION: A small proportion of patients presenting with SNHL tested positive on DBS. Of cCMV-positive children, most presented with profound hearing loss in the worse-hearing ear, and 50% of cCMV-positive children developed progressive hearing loss in the initially better-hearing ear. Prognostic information afforded by etiologic confirmation of cCMV infection informed decision-making concerning cochlear implantation in these cases.

Lee H, Lee H, Noh H.
Prediction of uptake and retention of conventional hearing aids in Korean pediatric patients with unilateral hearing loss.

Int J Pediatr Otorhinolaryngol. 2019 Jan;116:130-134. doi: 10.1016/j.ijporl.2018.10.037. Epub 2018 Oct 26.

OBJECTIVE: The purpose of this study was to describe and predict hearing aid uptake and retention in Korean pediatric patients with unilateral hearing loss (UHL) in a secondary referral hospital.

METHODS: This was a retrospective study using clinical data collected at the time of UHL diagnosis. The study included data collected from 2009 to 2016. Serial audiograms were extracted from clinical charts, and follow-up status and rehabilitation decisions were analyzed.

RESULTS: Of 102 children and adolescents (9.5 ± 5.1 years, 64 male), 52.9% followed a check-up schedule, and 31 (30.4%) obtained a hearing aid. Hearing threshold and speech discrimination scores were predictive parameters of hearing aid uptake. Among those who used a hearing aid, 17 (56.7%) subjects used it successfully based on significant predictive parameters of channel number.

CONCLUSION: Hearing aid retention in pediatric patients seems less predictable than in adults with UHL. No good predictable parameter for hearing aid retention was identified except channel number for pediatric UHL cases. Regular monitoring of hearing and selection of a multi-channel hearing aid are crucial to minimize the potential negative effects of UHL.

Lee JM, Nozu K, Choi DE, Kang HG, Ha IS, Cheong HI.

Features of Autosomal Recessive Alport Syndrome: A Systematic Review.

J Clin Med. 2019 Feb 3;8(2). pii: E178. doi: 10.3390/jcm8020178.

ABSTRACT: Alport syndrome (AS) is one of the most frequent hereditary nephritis leading to end-stage renal disease (ESRD). Although X-linked (XLAS) inheritance is the most common form, cases with autosomal recessive inheritance with mutations in *COL4A3* or *COL4A4* are being increasingly recognized. A systematic review was conducted on autosomal recessive Alport syndrome (ARAS). Electronic databases were searched using related terms (until Oct 10th, 2018). From 1601 articles searched, there were 26 eligible studies with 148 patients. Female and male patients were equally affected. About 62% of patients had ESRD, 64% had sensorineural hearing loss (SNHL) and 17% had ocular manifestation. The median at onset was 2.5 years for hematuria (HU), 21 years for ESRD, and 13 years for SNHL. Patients without missense mutations had more severe outcomes at earlier ages, while those who had one or two missense mutations had delayed onset and lower prevalence of extrarenal manifestations. Of 49 patients with kidney biopsy available for electron microscopy (EM) pathology, 42 (86%) had typical glomerular basement membrane (GBM) changes, while 5 (10%) patients showed GBM thinning only. SNHL developed earlier than previously reported. There was a genotype phenotype correlation according to the number of missense mutations. Patients with missense mutations had delayed onset of hematuria, ESRD, and SNHL and lower prevalence of extrarenal manifestations.

Leigh J, Farrell R, Courtenay D, Dowell R, Briggs R.

Relationship Between Objective and Behavioral Audiology for Young Children Being Assessed for Cochlear Implantation: Implications for CI Candidacy Assessment.

Otol Neurotol. 2019 Mar;40(3):e252-e259. doi: 10.1097/MAO.0000000000002125.

OBJECTIVE: This study aimed to evaluate the feasibility of making cochlear implant recommendations based on diagnostic ABR and ASSR results. The goal was to challenge the need for behavioral audiometry as part of the standard cochlear implant assessment battery for infants with profound hearing loss and to reduce the age at which cochlear implant recommendation was made.

STUDY DESIGN: A retrospective review of 123 patient files for children referred to the pediatric cochlear implant service before 3 years of age over a 3-year period was undertaken. Results for click-ABR, ASSR, and behavioral audiology at 500, 1k, 2k and 4k Hz, and tympanometry were collected and relationships were investigated for 64 children who met the inclusion criteria. Data were excluded for 59 children due to the presence of auditory neuropathy findings, middle ear pathology at the time of testing, if ASSR was not assessed at intensity levels >85dB, and/or behavioral testing was judged to be unreliable by two experienced clinicians.

SETTING: Primary care pediatric cochlear implant program located within a hospital setting.

PATIENTS: Pediatric patients referred for cochlear implant evaluation before 3 years of age.

INTERVENTIONS(S): Children were assessed using ABR, ASSR, and behavioral audiometry for identification and confirmation of hearing loss.

MAIN OUTCOME MEASURES(S): Correlation between diagnostic click-ABR and ASSR thresholds and subsequently obtained behavioral hearing thresholds.

RESULTS: Results for objective measures (click-ABR and ASSR) were significantly correlated with behavioral results. The correlations, however, were poorer than expected with limited predictive value. For 6 of the 64 children click-ABR and/or ASSR suggested profound hearing loss and corresponding behavioral hearing threshold was found to be in the severe hearing loss range.

CONCLUSIONS: Findings of this study do not support making cochlear implant recommendations based on the findings of diagnostic click-ABR and ASSR alone. Investigating ways to reduce the average age children with severe-to-profound hearing loss receive a cochlear implant is a priority for the study institution. An alternate

evaluation pathway for infants which incorporates a multifaceted assessment is warranted and will be the focus of future work at the study institution.

Li Y, Shen M, Long M.
A preliminary study of auditory mismatch response on the day of cochlear implant activation in children with hearing aids prior implantation.
PLoS One. 2019 Jan 7;14(1):e0210457. doi: 10.1371/journal.pone.0210457. eCollection 2019.

OBJECTIVE: The study aimed to explore the characteristics of auditory mismatch response (MMR) in hearing-impaired children on the day when the cochlear implant (CI) was started (power-up) and the speech processor was programmed, and to investigate the effects of wearing hearing aids (HAs) before cochlear implantation on the early stage of postoperative auditory cortex plasticity, providing some demonstrative data for the objective evaluation of postoperative early auditory ability in children who underwent cochlear implantation.
METHODS: The participants were 34 children with profound sensorineural hearing loss, who underwent cochlear implantation. The classical passive Oddball paradigm was adopted, using a pair of vowels which only have different lexical tones. The standard stimulus was /a2/ and the devious stimulus was /a4/.
RESULTS: 1) On the day of CI activation, the auditory MMR has been elicited in 30 children; the MMR incidence was 88%. 2) We observed both positive and negative auditory MMR waveforms. And logistic regression analysis showed that it was influenced by the age at cochlear implantation. 3) The duration with HA before surgery significantly influenced the MMR latency. The children with longer duration of HA use have much earlier latency of MMR. 4) There was a significant positive correlation between the age at HA use initiation and MMR amplitude. Earlier initial HA use was associated with smaller amplitude.
CONCLUSIONS: MMR in response to Mandarin lexical tone can be recorded in most pediatric patients who had experience with HA on the day of CI power up. MMR is closely associated with the age at cochlear implantation, duration of HA use, and the age at HA use initiation. Hearing-impaired children should wear HA as early as possible and ensure consistent usage.

Liu Y, Ye L, Zhu P, Wu J, Tan S, Chen J, Wu C, Zhong Y, Wang Y, Li X, Liu H.
Genetic screening involving 101 hot spots for neonates not passing newborn hearing screening and those random recruited in Dongguan.
Int J Pediatr Otorhinolaryngol. 2019 Feb;117:82-87. doi: 10.1016/j.ijporl.2018.11.008. Epub 2018 Nov 22.

ABSTRACT: In order to investigate essential molecular causes for hearing loss and mutation frequency of deafness-related genes, 1315 newborns who did not pass the Newborn Hearing Screening (NHS) (audio-no-pass) and 1000 random-selected infants were subjected to detection for 101 hotspot mutations in 18 common deafness-related genes. Totally, 23 alleles of 7 deafness genes were detected out. Significant difference ($\chi^2 = 25.320$, $p = 0.000$) existed in causative mutation frequency between audio-no-pass group (81/1315, 6.160%) and random-selected cohort (18/1000, 1.80%). Of the genes detected out, GJB2 gene mutation was with significant difference ($\chi^2 = 75.132$, $p = 0.000$) between audio-no-pass group (417/1315, 31.711%) and random-selected cohort (159/1000, 15.900%); c.109G > A was the most common allele, as well as the only one with significantly different allele frequency ($\chi^2 = 79.327$, $p = 0.000$) between audio-no-pass group (392/1315, 16.84%) and random-selected cohort (140/1000, 7.55%), which suggested c.109G > A mutation was critical for newborns' hearing loss. This study performed detection for such a large scale of deafness-associated genes and for the first time compared mutations between audio-no-pass and random-recruited neonates, which not only provided more reliable DNA diagnosis result for medical practioners and enhanced clinical care for the newborns, but gave more accurate estimation for mutation frequency.

Liu Y, Hu C, Liu C, Liu D, Mei L, He C, Jiang L, Wu H, Chen H, Feng Y.
A rapid improved multiplex ligation detection reaction method for the identification of gene mutations in hereditary hearing loss.
PLoS One. 2019 Apr 11;14(4):e0215212. doi: 10.1371/journal.pone.0215212. eCollection 2019.

ABSTRACT: Hearing loss (HL) is a common sensory disorder. More than half of HL cases can be attributed to genetic causes. There is no effective therapy for genetic HL at present, early diagnosis to reduce the incidence of genetic HL is important for clinical intervention in genetic HL. Previous studies have identified 111 nonsyndromic hearing loss genes. The most frequently mutated genes identified in NSHL patients in China include GJB2, SLC26A4, and the mitochondrial gene MT-RNR1. It is important to develop HL gene panels in Chinese population, which allow for etiologic diagnosis of both SHL and NSHL. In this study, a total of 220 unrelated Han Chinese patients with bilateral progressive SNHL and 50 unrelated healthy controls were performed Single nucleotide polymorphism (SNP) genotyping using an improved multiplex ligation detection reaction (iMLDR) technique, is to simultaneously detect a total of 32 mutations in ten HL genes, covering all currently characterized mutations involved in the etiology of nonsyndromic or syndromic hearing loss in the Chinese population. The 49 positive samples with known mutations were successfully detected using the iMLDR

Technique. For 171 SNHL patients, gene variants were found in 57 cases (33.33%), among which, 30 patients carried mutations in GJB2, 14 patients carried mutations in SLC26A4, seven patients carried mutations in GJB3, and six patients carried mutations in MT-RNR1. The molecular etiology of deafness was confirmed in 12.9% (22/171) of patients carried homozygous variants. These results were verified by Sanger sequencing, indicating that the sensitivity and specificity of the iMLDR technique was 100%. We believe that the implementation of this population-specific technology at an efficient clinical level would have great value in HL diagnosis and treatment.

Lu Y1, Zhou L, Imrit TS, Liu A.
Sudden Sensorineural Hearing Loss in Children: Clinical Characteristics, Etiology, Treatment Outcomes, and Prognostic Factors.
Otol Neurotol. 2019 Apr;40(4):446-453. doi: 10.1097/MAO.0000000000002190.

OBJECTIVE: To investigate the clinical characteristics, etiology, treatment outcomes, and prognostic factors of sudden sensorineural hearing loss (SSNHL) in children to guide the clinical diagnosis and treatment of SSNHL in the pediatric population.
STUDY DESIGN: Retrospective case review.
SETTING: Tertiary referral center.
PATIENTS: Patients diagnosed with SSNHL from November 2011 to December 2017 with relatively complete clinical data.
INTERVENTION: Diagnosis and systemic treatment of SSNHL.
MAIN OUTCOME MEASURES: Patients' clinical characteristics, etiology, laboratory tests, imaging, pure-tone audiometry at admission, and discharge were analyzed.
RESULTS: A total of 25 children and 149 adults with SSNHL were included. Recent or previous viral infection rates (81.8%) and fasting blood glucose level (5.23+1.47mmol/L) in children with SSNHL were lower than those in adult SSNHL patients (p=0.033, p=0.033). Autoimmune abnormalities (90.0%) and plasma fibrinogen abnormalities (27.3%) were higher in children with SSNHL than those in adult SSNHL patients (40.0%, 8.8%, respectively, p<0.05). The recovery rate in children (38.4%) with SSNHL is comparable to that in adults (22.6%), but children have a higher complete rate compared to adults (26.9%, 11.3%, respectively, p<0.05). Children with a profound audiometric curve had a worse prognosis in comparison to other types of audiometric curves (p=0.041).
CONCLUSIONS: Children with SSNHL have a lower rate of viral infection in comparison to adults with SSNHL. Fasting blood glucose levels, complement C3, C4, and fibrinogen may be closely related to childhood SSNHL. The recovery rate in children with SSNHL is comparable to that in adults, but children have a higher complete rate compared to adults. A profound hearing curve is an unfavorable prognostic factor in both children and adults with SSNHL.

Macielak RJ, Mattingly JK, Findlen UM, Moberly AC, Malhotra PS, Adunka OF.
Audiometric findings in children with unilateral enlarged vestibular aqueduct.
Int J Pediatr Otorhinolaryngol. 2019 May;120:25-29. doi: 10.1016/j.ijporl.2019.01.034. Epub 2019 Jan 25.

OBJECTIVE: To evaluate the prevalence of bilateral hearing loss in children with unilateral enlarged vestibular aqueduct (EVA) at a single institution.
METHODS: A retrospective case review was performed at a tertiary care pediatric referral center involving children with radiologic findings of unilateral EVA and normal labyrinthine anatomy of the contralateral ear diagnosed via CT and/or MRI. The main outcome measure of interest is the number of patients with unilateral EVA who were diagnosed with bilateral hearing loss.
RESULTS: Sixty-one pediatric patients were identified. The mean audiometric follow-up was 48.2 months (0-150). Three (4.9%) patients with unilateral EVA were noted to have bilateral hearing loss, and this rate was not significantly different (p = 1.0) from the rate reported in a comparison group of patients with contralateral hearing loss (6.0%) without an EVA. The pure-tone average (defined as the average dB HL at 500, 1000, 2000, and 4000 Hz) in the group with bilateral hearing loss was 31.3 dB HL in the better hearing ear and 79.6 dB HL in the worse hearing ear, with the difference being statistically significant (p = 0.02). In the unilateral EVA patients without contralateral hearing loss (n = 56, 91.8%), the PTA was 9.4 dB HL in the better hearing ear and 51.9 dB HL in the worse hearing ear, with the difference being statistically significant (p < 0.001). Two patients (3.3%) with unilateral EVA were found to have hearing within normal limits bilaterally. The EVA was ipsilateral to the worse hearing ear in all cases.
CONCLUSION: The prevalence of bilateral hearing loss in children with unilateral EVA appears to be low. Specifically, it may be no different than the rate of contralateral hearing loss in children with unilateral hearing loss without an EVA. The present report is somewhat different than the previously described prevalence in the literature. This difference could be related to the imaging type and diagnostic criteria used, the patients included, the source of the identified patents, and the overall population of patients studied.

Maluleke NP, Khoza-Shangase K, Kanji A.
Communication and school readiness abilities of children with hearing impairment in South Africa: A retrospective review of early intervention preschool records.
S Afr J Commun Disord. 2019 Feb 28;66(1):e1-e7. doi: 10.4102/sajcd.v66i1.604.

BACKGROUND: The national prevalence of hearing impairment in South Africa is estimated to be four to six in every 1000 live births in the public health care sector. An undetected hearing impairment in childhood can lead to delayed speech and language development as well as put the child at risk of not achieving the necessary school readiness abilities that will enable them to achieve academic success. However, through early hearing detection and intervention services, children with hearing impairment can develop communication and school readiness abilities on par with children with normal hearing.
OBJECTIVE: The aim of the study was to describe communication and school readiness abilities of children who were identified with hearing impairment and enrolled in early intervention (EI) preschools in Gauteng.
METHODS: Within a descriptive research study design, a retrospective record review was conducted on files of eight children, ranging in age from 9 years and 7 months to 12 years and 7 months, identified with a hearing impairment and enrolled in EI preschools in Gauteng, South Africa. Descriptive statistics were used to analyse the data, using frequency distribution and measures of central tendency.
RESULTS: Current findings revealed that children with hearing impairment who were enrolled in EI preschools in Gauteng were identified late. This consequently led to delayed ages at initiation of EI services when compared to international benchmarks and the Health Professions Council of South Africa›s (HPCSA) guidelines of 2018. Consequently, participants presented with below average communication and school readiness abilities, which are characteristic of hearing impairment that is identified late.
CONCLUSIONS: Transference of current contextually relevant research findings into practice by both the Department of Health and the Department of Basic Education forms part of future directions from this study. This conversion of research findings into service delivery must be conducted in a systematic manner at all levels in these two sectors to facilitate achievement of Early Hearing Detection and Intervention (EHDI), resulting in better communication and school readiness outcomes.

Manjaly JG, Nash R, Ellis W, Britz A, Lavy JA, Shaida A, Saeed SR, Khalil SS.
Hearing Preservation With Standard Length Electrodes in Pediatric Cochlear Implantation.
Otol Neurotol. 2018 Oct;39(9):1109-1114. doi: 10.1097/MAO.0000000000001917.

OBJECTIVE: Preserving low frequencies following cochlear implantation improves outcomes and allows patients to use a combination of electrical and acoustic stimulation. This importance has been reflected in advances in electrode design and refined surgical techniques. Full insertion of standard length electrodes may be advantageous over shortened electrodes because more electrodes can be activated over time if low frequency hearing loss progresses. Surgeons must counsel patients over this choice but data is lacking regarding the degree and likelihood of hearing preservation achievable with standard length electrodes in children. We report our experience using standard length cochlear implant arrays for hearing preservation in children.
METHODS: Retrospective case series.
INCLUSION CRITERIA: preoperative hearing ≤85dB HL at 250Hz and aged ≤18 years. Hearing preservation percentages are calculated using the HEARRING group formula. (Equation is included in full-text article.) Preservation of > 75% was considered complete, 25 to 75% partial, and 1 to 25% minimal. Patients were implanted with either MED-EL FLEX28 or Cochlear Nucleus CI522. Standardized operative technique with facial recess approach, posterior tympanotomy and minimally traumatic round window insertion.
RESULTS: Fifty-two implantations in 27 pediatric patients met inclusion criteria. Mean age at implantation: 9.8 years. Average latest audiogram: 8 months. Mean total pre- and postoperative pure-tone averages were 82.8 and 92.6dB. Seventeen (33%) ears demonstrated complete hearing preservation, 22 (42%) ears partial hearing preservation, 7 (13%) minimal hearing preservation, and 6 (12%) exhibited no acoustic hearing postoperatively. Mean hearing preservation was 55.5%.
CONCLUSION: Hearing preservation is achievable to varying degrees in pediatric cochlear implantation using standard length electrodes though it is difficult to predict preoperatively which children may benefit. This study is among the largest additions to the knowledge base for this patient group.

Mauldin L.
Don't look at it as a miracle cure: Contested notions of success and failure in family narratives of pediatric cochlear implantation.
Soc Sci Med. 2019 May;228:117-125. doi: 10.1016/j.socscimed.2019.03.021. Epub 2019 Mar 16.

ABSTRACT: Cochlear implants (CIs) are a routine treatment for children identified with a qualifying hearing loss. The CI, however, must be accompanied by a long-term and intense auditory training regimen in order to possibly acquire spoken language with the device. This research investigates families' experiences when they opted for the CI and undertook the task of auditory training, but the child failed to achieve what might be clinically

considered “success” - the ability to function solely using spoken language. Using a science and technology studies informed approach that places the CI within a complex sociotechnical system, this research shows the uncertain trajectory of the CI, as well as the contingency of the very notions of success and failure. To do so, data from in-depth interviews with a diverse sample of parents (n = 11) were collected. Results show the shifting definitions of failure and success within families, as well as suggest areas for further exploration regarding clinical practice and pediatric CIs. First, professionals’ messaging often conveyed to parents a belief in the infallibility of the CI, this potentially caused “soft failure” to go undetected and unmitigated. Second, speech assessments used in clinical measurements of outcomes did not capture a holistic understanding of a child’s identity and social integration, leaving out an important component for consideration of what a ‘good outcome’ is. Third, minority parents experience structural racism and clinical attitudes that may render “failure” more likely to be identified and expected in these children, an individualizing process that allows structural failures to go uncritiqued.

McCrary H, Sheng X, Greene T, Park A.
Long-term hearing outcomes of children with symptomatic congenital CMV treated with valganciclovir.
Int J Pediatr Otorhinolaryngol. 2019 Mar;118:124-127. doi: 10.1016/j.ijporl.2018.12.027. Epub 2018 Dec 21.

OBJECTIVES: Congenital human cytomegalovirus (cCMV) is a leading cause of pediatric hearing loss. Recent literature has suggested that valganciclovir (VGCV) therapy can improve hearing outcomes. The objective of this study was to evaluate the long-term hearing outcomes among symptomatic CMV patients treated with VGCV.
METHODS: A retrospective chart review of symptomatic CMV patients treated with VGCV was completed. The primary endpoint was the change in best ear hearing scores prior to treatment and after follow-up audiograms. A paired-sample t-test was used to evaluate the data.
RESULTS: A total of 16 children were included in the study and participants were followed for an average of 3.2 years. There was a measurable worsening, but not a statistically significant change in the best ear hearing scores, where the mean change was 11.9 dB (p-value = 0.070). However, 14/16 patients (87.5%, p-value<0.001) were found to have clinically significant worsening of hearing. The mean change in hearing scores for the left and right ear was 14.2 dB (p-value = 0.023) and 15.5 dB (p-value = 0.032), respectively. Mean elapsed time for progressive loss was 2.6 ± 0.2 years. When comparing the better or worse ear, there was no pattern for which ear deteriorated earlier or more frequently.
CONCLUSIONS: Our data did show a measurable, but not a statistically significant worsening outcome in best ear hearing. There was a significant change in both left and right ear hearing. Our results suggest that VGCV may provide only a short-term improvement in hearing outcomes; however, these preliminary post-hoc findings suggest the need for a more rigorous evaluation.

McDaniel J, Camarata S, Yoder P.
Comparing Auditory-Only and Audiovisual Word Learning for Children With Hearing Loss.
J Deaf Stud Deaf Educ. 2018 Oct 1;23(4):382-398. doi: 10.1093/deafed/eny016.

ABSTRACT: Although reducing visual input to emphasize auditory cues is a common practice in pediatric auditory (re)habilitation, the extant literature offers minimal empirical evidence for whether unisensory auditory-only (AO) or multisensory audiovisual (AV) input is more beneficial to children with hearing loss for developing spoken language skills. Using an adapted alternating treatments single case research design, we evaluated the effectiveness and efficiency of a receptive word learning intervention with and without access to visual speechreading cues. Four preschool children with prelingual hearing loss participated. Based on probes without visual cues, three participants demonstrated strong evidence for learning in the AO and AV conditions relative to a control (no-teaching) condition. No participants demonstrated a differential rate of learning between AO and AV conditions. Neither an inhibitory effect predicted by a unisensory theory nor a beneficial effect predicted by a multisensory theory for providing visual cues was identified. Clinical implications are discussed.

McKearney RM, MacKinnon RC.
Objective auditory brainstem response classification using machine learning.
Int J Audiol. 2019 Apr;58(4):224-230. doi: 10.1080/14992027.2018.1551633. Epub 2019 Jan 21.

OBJECTIVE: The objective of this study was to use machine learning in the form of a deep neural network to objectively classify paired auditory brainstem response waveforms into either: ‘clear response’, ‘inconclusive’ or ‘response absent’.
DESIGN: A deep convolutional neural network was constructed and fine-tuned using stratified 10-fold cross-validation on 190 paired ABR waveforms. The final model was evaluated on a test set of 42 paired waveforms.
STUDY SAMPLE: The full dataset comprised 232 paired ABR waveforms recorded from eight normal-hearing individuals. The dataset was obtained from the PhysioBank database. The paired waveforms were independently labelled by two audiological scientists in order to train the network and evaluate its performance.
RESULTS: The trained neural network was able to classify paired ABR waveforms with 92.9% accuracy. The

sensitivity and the specificity were 92.9% and 96.4%, respectively.
CONCLUSIONS: This neural network may have clinical utility in assisting clinicians with waveform classification for the purpose of hearing threshold estimation. Further evaluation using a large clinically obtained dataset would provide further validation with regard to the clinical potential of the neural network in diagnostic adult testing, newborn testing and in automated newborn hearing screening.

Mishra S, Pandey H, Srivastava P, Mandal K, Phadke SR.
Connexin 26 (GJB2) Mutations Associated with Non-Syndromic Hearing Loss (NSHL).
Indian J Pediatr. 2018 Dec;85(12):1061-1066. doi: 10.1007/s12098-018-2654-8. Epub 2018 Mar 15.
OBJECTIVE: To determine the prevalence and spectrum of Connexin 26 (GJB2) mutations in pre-lingual non-syndromic hearing loss (NSHL) patients in authors' centre and to review the data of Indian patients from the literature.
METHODS: Sanger sequencing of entire coding region contained in single exon (Exon 2) of GJB2 gene in 15 patients of NSHL.
RESULTS: GJB2 mutations were found in 40% (6/15) of NSHL patients, out of which mono-allelic were 33.3% (2/6). Bi-allelic GJB2 mutations were identified in 4 of 6 patients. Most common GJB2 mutation identified was c.71G>A(p.W24X), comprising 30% of the total GJB2 mutant alleles. Six studies involving 1119 patients with NSHL were reviewed and 4 of them have reported c.71G>A(p.W24X) as the commonest mutation while 2 studies found c.35delG as the commonest. GJB2 mutations accounted for 10.9%-36% cases of NSHL. Sixteen other mutations in GJB2 gene were reported in Indian patients out of which 6 mutations other than c.71G>A(p.W24X) viz., c.35delG, c.1A>G(p.M1V), c.127G>A(p.V43 M), c.204C>G(p.Y86X), c.231G>A(p.W77X) and c.439G>A(p.E147K) were identified in the present study.
CONCLUSIONS: Connexin 26 (GJB2) mutations are responsible for 19.4% of NSHL in Indian population. The c.71G>A(W24X) and c.35delG were the most prevalent GJB2 mutations accounting for 72.2% (234 of 324 total mutated alleles from 7 studies) and 15.4% (50 of 324 total mutated alleles from 7 studies) respectively. Thus, screening of these two common mutations in GJB2 gene by polymerase chain reaction and restriction fragment length polymorphism (PCR-RFLP) would greatly help in providing easy genetic diagnosis and help in genetic counseling of the families with NSHL.

Muñoz K, Price T, Nelson L, Twohig M.
Counseling in Pediatric Audiology: Audiologists' Perceptions, Confidence, and Training.
J Am Acad Audiol. 2019 Jan;30(1):66-77. doi: 10.3766/jaaa.17087. Epub 2017 Dec 21.
BACKGROUND: Pediatric audiologists are an important source of support for parents when a child is identified with hearing loss. As parents learn how to manage their child's hearing loss they often need help navigating challenges that arise; however, audiologists may experience a variety of barriers implementing effective counseling strategies. Many internal and external barriers experienced by parents can be appropriately supported and navigated within audiology services.
PURPOSE: To investigate audiologists' perceptions, training, and confidence related to counseling and to explore the influence of years practicing audiology and taking a counseling course on perceptions and confidence.
RESEARCH DESIGN: A cross-sectional, population-based survey.
STUDY SAMPLE: Three hundred and fifty surveys were analyzed from pediatric audiologists across the U.S. Responses were received from 26 states and one U.S. territory.
DATA COLLECTION AND ANALYSIS: Data were collected through the mail and online. Descriptive and comparative statistics were used to analyze the information. Content analysis was performed to identify emergent themes from the responses to open-ended questions.
RESULTS: Pediatric audiologists reported their perceptions about importance of counseling skills, challenges they encounter, their confidence in counseling, and how often they use the skills when needed in practice. Most audiologists (≥75%) felt it was very or extremely important to talk with parents about nine of the ten items (e.g., their [parents'] expectations). Three-fourth of the audiologists reported experiencing a moderate challenge or greater in knowing how to assess the presence of psychosocial challenges and in having enough time to address emotional needs. Many of the audiologists felt very or extremely confident in guiding parents in the development of an action plan (62%) and determining if parents have external barriers (60%). Approximately one-third or less of the participants reported performing any of the skills (e.g., determining if parent has external or internal barriers) ≥75% of the time, and a statistically significant difference was found with participants practicing ≤10 yr using the skills more frequently than participants practicing for ≥11 yr. In addition, there was a statistically significant difference between participants who had taken an audiology-specific counseling course and those who had not; those who had reported being more confident and using counseling skills more often than audiologists did not have a counseling course.
CONCLUSIONS: This study found strong support for audiologist perceived importance of counseling; however,

fewer audiologists reported confidence in their counseling skills and in using counseling skills. Counseling training was variable; audiologists would benefit from a more systematic approach to counseling instruction within graduate training.

Myers J, Kei J, Aithal S, Aithal V, Driscoll C, Khan A, Manuel A, Joseph A, Malicka AN.
Development of a Diagnostic Prediction Model for Conductive Conditions in Neonates Using Wideband Acoustic Immittance.
Ear Hear. 2018 Nov/Dec;39(6):1116-1135. doi: 10.1097/AUD.0000000000000565.

OBJECTIVES: Wideband acoustic immittance (WAI) is an emerging test of middle-ear function with potential applications for neonates in screening and diagnostic settings. Previous large-scale diagnostic accuracy studies have assessed the performance of WAI against evoked otoacoustic emissions, but further research is needed using a more stringent reference standard. Research into suitable quantitative techniques to analyze the large volume of data produced by WAI is still in its infancy. Prediction models are an attractive method for analysis of multivariate data because they provide individualized probabilities that a subject has the condition. A clinically useful prediction model must accurately discriminate between normal and abnormal cases and be well calibrated (i.e., give accurate predictions). The present study aimed to develop a diagnostic prediction model for detecting conductive conditions in neonates using WAI. A stringent reference standard was created by combining results of high-frequency tympanometry and distortion product otoacoustic emissions.
DESIGN: High-frequency tympanometry and distortion product otoacoustic emissions were performed on both ears of 629 healthy neonates to assess outer- and middle-ear function. Wideband absorbance and complex admittance (magnitude and phase) were measured at frequencies ranging from 226 to 8000 Hz in each neonate at ambient pressure using a click stimulus. Results from one ear of each neonate were used to develop the prediction model. WAI results were used as logistic regression predictors to model the probability that an ear had outer/middle-ear dysfunction. WAI variables were modeled both linearly and nonlinearly, to test whether allowing nonlinearity improved model fit and thus calibration. The best-fitting model was validated using the opposite ears and with bootstrap resampling.
RESULTS: The best-fitting model used absorbance at 1000 and 2000 Hz, admittance magnitude at 1000 and 2000 Hz, and admittance phase at 1000 and 4000 Hz modeled as nonlinear variables. The model accurately discriminated between normal and abnormal ears, with an area under the receiver-operating characteristic curve (AUC) of 0.88. It effectively generalized to the opposite ears (AUC = 0.90) and with bootstrap resampling (AUC = 0.85). The model was well calibrated, with predicted probabilities aligning closely to observed results.
CONCLUSIONS: The developed prediction model accurately discriminated between normal and dysfunctional ears and was well calibrated. The model has potential applications in screening or diagnostic contexts. In a screening context, probabilities could be used to set a referral threshold that is intuitive, easy to apply, and sensitive to the costs associated with true- and false-positive referrals. In a clinical setting, using predicted probabilities in conjunction with graphical displays of WAI could be used for individualized diagnoses. Future research investigating the use of the model in diagnostic or screening settings is warranted.

Nada DW, El Khouly RM, Gadow SE, Hablas SA, Aboelhawa MA, Al Ashkar DS, El Barbary AM, Hussein MS, Rageh E, Elsalawy AM, Abo-Zaid MH, Elshweikh S, El Gharib AM.
The role of auditory evoked potentials and otoacoustic emissions in early detection of hearing abnormalities in Behçet's disease patients. A case control study.
Clin Exp Rheumatol. 2018 Nov-Dec;36(6 Suppl 115):45-52. Epub 2018 May 10.

OBJECTIVES: To determine the types and to assess the role of auditory evoked potentials and otoacoustic emissions in early detection of hearing abnormalities in Behçet's disease (BD) patients. Their correlations with disease activity were also considered.
METHODS: Thirty patients with BD and thirty apparently sex- and age-matched healthy volunteers were included in this study. Auditory evaluation included pure tone audiometry (PTA), otoacoustic emissions (TEOAEs, DPOAE), auditory brainstem response test (ABR) and cortical auditory evoked potentials (tone and speech CAEPs) for all patients and control.
RESULTS: The highest abnormality of CAEP latencies elicited by (500Hz and 1000 Hz) as well as speech stimuli (da and ga) among our BD patients was delayed P1 and N1 waves at 80 dB with greater bilateral affection, as well as significant differences between patients and controls. All our BD patients had a smaller amplitude of distortion product OAE (DPOAE) and S/N ratio at 1, 2, 4, 6 kHZ compared with controls and the differences were highly statistically significant (p=0.0001).
CONCLUSIONS: Being one of the autoimmune inner ear diseases (AIED), BD has a definite hearing impairment, even in the presence of normal hearing sensitivity, as evidenced by PTA. BD patients had a sub-clinical cochlear pathology which was not affected by disease activity or different organ affection. DPOAE (S/N ratio) proved to be a sensitive test in detecting minimal changes in cochlear pathology and the latencies of CAEPs

(tone and speech) measures were considered as sensitive indicators (100%) of early detection of hearing impairment in BD patients.

Nam GS, Kwak SH, Bae SH, Kim SH, Jung J, Choi JY.
Hyperbilirubinemia and Follow-up Auditory Brainstem Responses in Preterm Infants.
Clin Exp Otorhinolaryngol. 2019 May;12(2):163-168. doi: 10.21053/ceo.2018.00899. Epub 2018 Nov 9.

OBJECTIVES: Neonatal hyperbilirubinemia is considered one of the most common causative factors of hearing loss. Preterm infants are more vulnerable to neuronal damage caused by hyperbilirubinemia. This study aimed to evaluate the effect of hyperbilirubinemia on hearing threshold and auditory pathway in preterm infants by serial auditory brainstem response (ABR). In addition, we evaluate the usefulness of the unconjugated bilirubin (UCB) level compared with total serum bilirubin (TSB) on bilirubin-induced hearing loss.
METHODS: This study was conducted on 70 preterm infants with hyperbilirubinemia who failed universal newborn hearing screening by automated ABR. The diagnostic ABR was performed within 3 months after birth. Follow-up ABR was conducted in patients with abnormal results (30 cases). TSB and UCB concentration were compared according to hearing threshold by ABR.
RESULTS: The initial and maximal measured UCB concentration for the preterm infants of diagnostic ABR ≥40 dB nHL group (n=30) were statistically higher compared with ABR ≤35 dB nHL group (n=40) (P=0.031 and P=0.003, respectively). In follow-up ABR examination, 13 of the ABR ≥40 dB nHL group showed complete recovery, but 17 had no change or worsened. There was no difference in bilirubin level between the recovery group and non-recovery group.
CONCLUSION: UCB is a better predictor of bilirubin-induced hearing loss than TSB in preterm infants as evaluated by serial ABR. Serial ABR testing can be a useful, noninvasive methods to evaluate early reversible bilirubin-induced hearing loss in preterm infants.

Neumann K, Thomas JP, Voelter C, Dazert S.
A new adhesive bone conduction hearing system effectively treats conductive hearing loss in children.
Int J Pediatr Otorhinolaryngol. 2019 Apr 3;122:117-125. doi: 10.1016/j.ijporl.2019.03.014. [Epub ahead of print]

OBJECTIVES: Bone conduction hearing devices integrated in softbands (BCDSs) are frequently not well accepted by children with conductive hearing loss due to pressure on the head, sweating, or cosmetic stigma. A non-surgical hearing system (ADHEAR) uses a new bone conduction concept consisting of an audio processor connected to an adhesive adapter fixed behind the ear. This study is the first to evaluate the audiological and clinical outcome of this novel system, comparing it with conventional BCDSs in a short- and mid-term follow-up in children under 10 years of age.
METHODS: The ADHEAR was compared to a BCDS in 10 children with conductive hearing loss (age: 0.7-9.7 years). Aided and unaided pure tone/behavioral observational audiometry and, if applicable, speech audiometry in quiet and noise were performed initially with both devices and after 8 weeks with the ADHEAR alone. The subjective hearing gain and usage of the new hearing system, as well as patients' and parents' satisfaction were assessed using questionnaires.
RESULTS: The functional gain with the ADHEAR averaged over 0.5, 1, 2, and 4 kHz exceeded that of the conventional BCDS (35.6 dB ± 15.1 vs. 29.9 dB ± 14.6, p = .001, n = 9 ears). Speech perception in quiet and noise (n = 8) improved in the aided situation similarly for both hearing devices. The parents of 8 of 10 children evaluated the ADHEAR system as being useful. Minor wearing problems occurred occasionally. Eight children continued using the ADHEAR after the study, one received an active middle ear implant and one continued to use a BCDS.
CONCLUSION: The ADHEAR system is a promising solution for children with conductive hearing loss or chronically draining ears.

Ngui LX, Tang IP, Prepageran N, Lai ZW.
Comparison of distortion product otoacoustic emission (DPOAE) and automated auditory brainstem response (AABR) for neonatal hearing screening in a hospital with high delivery rate.
Int J Pediatr Otorhinolaryngol. 2019 May;120:184-188. doi: 10.1016/j.ijporl.2019.02.045. Epub 2019 Feb 27.

INTRODUCTION: Congenital hearing loss is one of the commonest congenital anomalies. Neonatal hearing screening aims to detect congenital hearing loss early and provide prompt intervention for better speech and language development. The two recommended methods for neonatal hearing screening are otoacoustic emission (OAE) and automated auditory brainstem response (AABR).
OBJECTIVE: To study the effectiveness of distortion product otoacoustic emission (DPOAE) and automated auditory brainstem response (AABR) as first screening tool among non-risk newborns in a hospital with high delivery rate.
METHOD: A total of 722 non-risk newborns (1444 ears) were screened with both DPOAE and AABR prior to discharge within one month. Babies who failed AABR were rescreened with AABR ± diagnostic auditory

brainstem response tests within one month of age.
RESULTS: The pass rate for AABR (67.9%) was higher than DPOAE (50.1%). Both DPOAE and AABR pass rates improved significantly with increasing age (p-value<0.001). The highest pass rate for both DPOAE and AABR were between the age of 36-48 h, 73.1% and 84.2% respectively. The mean testing time for AABR (13.54 min ± 7.47) was significantly longer than DPOAE (3.52 min ± 1.87), with a p-value of <0.001.
CONCLUSIONS: OAE test is faster and easier than AABR, but with higher false positive rate. The most ideal hearing screening protocol should be tailored according to different centre.

Nunes ADDS, Silva CRL, Balen SA, Souza DLB, Barbosa IR.
Prevalence of hearing impairment and associated factors in school-aged children and adolescents: a systematic review.
Braz J Otorhinolaryngol. 2019 Mar - Apr;85(2):244-253. doi: 10.1016/j.bjorl.2018.10.009. Epub 2018 Dec 1.

INTRODUCTION: Hearing impairment is one of the communication disorders of the 21st century, constituting a public health issue as it affects communication, academic success, and life quality of students. Most cases of hearing loss before 15 years of age are avoidable, and early detection can help prevent academic delays and minimize other consequences.
OBJECTIVE: This study researched scientific literature for the prevalence of hearing impairment in school-aged children and adolescents, with its associated factors. This was accomplished by asking the defining question: "What is the prevalence of hearing impairment and its associated factors in school-aged children and adolescents?"
METHODS: Research included the databases PubMed/MEDLINE, LILACS, Web of Science, Scopus and SciELO, and was carried out by two researchers, independently. The selected papers were analyzed on the basis of the checklist provided by the report Strengthening the Reporting of Observational Studies in Epidemiology.
RESULTS: From the 463 papers analyzed, 26 fulfilled the criteria and were included in the review presented herein. The detection methods, as well as prevalence and associated factors, varied across studies. The prevalence reported by the studies varied between 0.88% and 46.70%. Otologic and non-otologic factors were associated with hearing impairment, such as middle ear and air passage infections, neo- and post-natal icterus, accumulation of cerumen, family history, suspicion of parents, use of earphones, age and income.
CONCLUSION: There is heterogeneity regarding methodology, normality criteria, and prevalence and risk factors of studies about hearing loss in adolescents and school-aged children. Nevertheless, the relevance of the subject and the necessity of early interventions are unanimous across studies.

Núñez-Batalla F, Jáudenes-Casaubón C, Sequí-Canet JM, Vivanco-Allende A, Zubicaray-Ugarteche J.
Early diagnosis and treatment of unilateral or asymmetrical hearing loss in children: CODEPEH recommendations.
Acta Otorrinolaringol Esp. 2018 Dec 19. pii: S0001-6519(18)30178-X. doi: 10.1016/j.otorri.2018.09.004. [Epub ahead of print]

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

Olarte M, Bermúdez Rey MC, Beltran AP, Guerrero D, Suárez-Obando F, López G, García M, Ospina JC, Fonseca C, Bertolotto AM, Aldana N, Gelvez N, Tamayo ML.
Detection of hearing loss in newborns: Definition of a screening strategy in Bogotá, Colombia.
Int J Pediatr Otorhinolaryngol. 2019 Mar 26;122:76-81. doi: 10.1016/j.ijporl.2019.03.016. [Epub ahead of print]

OBJECTIVE: To describe the results from the hearing screening protocol adopted in a Hospital in Colombia emphasizing the importance of performing screening on an outpatient basis, when the newborn is more than 24 h old.
METHODS: A prospective study at Hospital Universitario San Ignacio in Bogota, Colombia was carried out, from May 1st, 2016 to Nov 30th, 2017, the study sample included 2.088 newborns examined using transient otoacoustic emissions.
RESULTS: We obtained written consent from the parents of 1.523 newborns and 24 individuals (1.6%) failed the first stage of the screening, nine cases unilateral and 15 bilateral. A total of nine neonates (0,6%) failed the second screening test, six cases unilateral and three bilateral. Four (0,3%) did not return to the second test. Our false altered screening rate was 0.7%.

CONCLUSIONS: In a developing country with limited human and economic resources, in which newborn early discharge is the norm, a newborn hearing screening program linked to infants' check-ups, that uses otoacoustic emissions after 48 h of life, seems a feasible option compare to the standard US protocol aiming to conduct hearing screening prior to discharge.

Palma S, Roversi MF, Bettini M, Mazzoni S, Pietrosemoli P, Lucaccioni L, Berardi A, Genovese E.
Hearing loss in children with congenital cytomegalovirus infection: an 11-year retrospective study based on laboratory database of a tertiary paediatric hospital.
Acta Otorhinolaryngol Ital. 2019 Feb;39(1):40-45. doi: 10.14639/0392-100X-2020.

ABSTRACT: Congenital cytomegalovirus infection is considered the main cause of infantile non-genetic neurosensory hearing loss. Although this correlation was described more than 50 years ago, the natural history of internal ear involvement has not yet been fully defined. Hearing loss is the most frequent sequela and is seen in a variable percentage up to 30%; the hearing threshold is characterised by fluctuations or progressive deterioration. The purpose of this study was to evaluate the prevalence of hearing loss in cases of congenital CMV infection from Modena county, starting from the database of the microbiology and virology reference laboratory. All children undergoing urine testing for suspected CMV infection or viral DNA testing on Guthrie Card in the period between January 2004 and December 2014 were enrolled in the study. Family paediatricians were contacted and asked about clinical information on the possible presence at birth or subsequent occurrence of hearing loss, excluding cases where this was not possible. The results showed an annual prevalence of congenital cytomegalovirus infection among suspected cases that was stable over time despite the progressive increase in subjects tested. The prevalence of hearing loss was in line with the literature, whereas in long-term follow-up cases of moderate, medium-to-severe hearing loss with late onset were not detected. The introduction of newborn hearing screening in the county has allowed early diagnosis of hearing loss at birth as non-TEOAE-born births underwent a urine virus test. Moreover, despite all the limitations of the study, we can conclude that European epidemiological studies are needed to better define the relationship between congenital CMV infection and internal ear disease as the impact of environmental and genetic factors is still not entirely clarified.

Pasternak Y, Attias J, Ely N, Amir J, Bilavsky E.
No risk factors for late onset hearing loss in asymptomatic congenital cytomegalovirus infants - close monitoring is needed.
Acta Paediatr. 2019 Apr 12. doi: 10.1111/apa.14814. [Epub ahead of print]

ABSTRACT: Congenital cytomegalovirus (cCMV) is the leading cause of congenital infections, affecting approximately 0.7% of live births worldwide. Although, 85%-90% of infected children are asymptomatic at birth, 10%-15% will develop late onset hearing impairment (1). The appropriate management of cCMV is controversial and data are needed to estimate the cost-effectiveness of universal versus targeted newborn screening. Many risk factors for late onset hearing deterioration have been proposed, but not confirmed.

Pourreza MR, Mohammadi H, Sadeghian L, Asgharzadeh S, Sehhati M, Tabatabaiefar MA.
Applying Two Different Bioinformatic Approaches to Discover Novel Genes Associated with Hereditary Hearing Loss via Whole-Exome Sequencing: ENDEAVOUR and HomozygosityMapper.
Adv Biomed Res. 2018 Oct 31;7:141. doi: 10.4103/abr.abr_80_18. eCollection 2018.

BACKGROUND: Hearing loss (HL) is a highly prevalent heterogeneous deficiency of sensory-neural system with involvement of several dozen genes. Whole-exome sequencing (WES) is capable of discovering known and novel genes involved with HL.
MATERIALS AND METHODS: Two pedigrees with HL background from Khuzestan province of Iran were selected. Polymerase chain reaction-sequencing of *GJB2* and homozygosity mapping of 16 DFNB loci were performed. One patient of the first and two affected individuals from the second pedigree were subjected to WES. The result files were analyzed using tools on Ubuntu 16.04. Short reads were mapped to reference genome (hg19, NCBI Build 37). Sorting and duplication removals were done. Variants were obtained and annotated by an online software tool. Variant filtration was performed. In the first family, ENDEAVOUR was applied to prioritize candidate genes. In the second family, a combination of shared variants, homozygosity mapping, and gene expression were implemented to launch the disease-causing gene.
RESULTS: *GJB2* sequencing and linkage analysis established no homozygosity-by-descent at any DFNB loci. Utilizing ENDEAVOUR, *BBX*: C.C857G (*P*.A286G), and *MYH15*: C.C5557T (*P*.R1853C) were put forward, but none of the variants co-segregated with the phenotype. Two genes, *UNC13B* and *TRAK1*, were prioritized in the homozygous regions detected by HomozygosityMapper.
CONCLUSION: WES is regarded a powerful approach to discover molecular etiology of Mendelian inherited disorders, but as it fails to enrich GC-rich regions, incapability of capturing noncoding regulatory regions and limited specificity and accuracy of copy number variations detection tools from exome data, it is assumed an

insufficient procedure.

Puia-Dumitrescu M, Bretzius OM, Brown N, Fitz-Henley JA, Ssengonzi R, Wechsler CS, Gray KD, Benjamin DK Sr, Smith PB, Clark RH, Gonzalez D, Hornik CP.
Evaluation of Gentamicin Exposure in the Neonatal Intensive Care Unit and Hearing Function at Discharge.
J Pediatr. 2018 Dec;203:131-136. doi: 10.1016/j.jpeds.2018.07.101. Epub 2018 Sep 21.

OBJECTIVE: To characterize the association between gentamicin dosing, duration of treatment, and ototoxicity in hospitalized infants.
STUDY DESIGN: This retrospective cohort study conducted at 330 neonatal intensive care units (2002-2014) included inborn infants exposed to gentamicin with available hearing screen results, and excluded infants with incomplete dosing data and major congenital anomalies. Our primary outcome was the final hearing screen result performed during hospitalization: abnormal (failed or referred for further testing in one or both ears) or normal (bilateral passed). The 4 measures of gentamicin exposure were highest daily dose, average daily dose, cumulative dose, and cumulative duration of exposure. We fitted separate multivariable logistic regression models adjusted for demographics, comorbidities, and other clinical events.
RESULTS: A total of 84 808 infants met inclusion/exclusion criteria; median (25th, 75th percentile) gestational age and birth weight were 35 weeks (33, 38) and 2480 g (1890, 3184), respectively. Failed hearing screens occurred in 3238 (3.8%) infants; failed screens were more likely in infants of lower gestational age and birth weight, who had longer hospital lengths of stay, higher rates of morbidities, and were small for gestational age. Median highest daily dose, average daily dose, and cumulative dose were 4.0 mg/kg/day (3.0, 4.0), 3.8 mg/kg/day (3.0, 4.0), and 12.1 mg/kg (9.1, 20.5), respectively. Median cumulative duration of exposure was 3 days (3, 6). In adjusted analysis, gentamicin dose and duration of therapy were not associated with hearing screen failure.
CONCLUSIONS: Gentamicin dosing and duration of treatment were not associated with increased odds of failed hearing screen at the time of discharge from initial neonatal intensive care unit stay.

Ramkumar V, Nagarajan R, Shankarnarayan VC, Kumaravelu S, Hall JW.
Implementation and evaluation of a rural community-based pediatric hearing screening program integrating in-person and tele-diagnostic auditory brainstem response (ABR).
BMC Health Serv Res. 2019 Jan 3;19(1):1. doi: 10.1186/s12913-018-3827-x.

BACKGROUND: In an attempt to reach remote rural areas, this study explores a community-based, pediatric hearing screening program in villages, integrating two models of diagnostic ABR testing; one using a tele-medicine approach and the other a traditional in-person testing at a tertiary care hospital.
METHODS: Village health workers (VHWs) underwent a five day training program on conducting Distortion Product Oto Acoustic Emissions (DPOAE) screening and assisting in tele-ABR. VHWs conducted DPOAE screening in 91 villages and hamlets in two administrative units (blocks) of a district in South India. A two-step DPOAE screening was carried out by VHWs in the homes of infants and children under five years of age in the selected villages. Those with 'refer' results in 2nd screening were recommended for a follow-up diagnostic ABR testing in person (Group A) at the tertiary care hospital or via tele-medicine (Group B). The overall outcome of the community-based hearing screening program was analyzed with respect to coverage, refer rate, follow-up rate for 2nd screenings and diagnostic testing. A comparison of the outcomes of tele-versus in-person diagnostic ABR follow-up was carried out.
RESULTS: Six VHWs who fulfilled the post training evaluation criteria were recruited for the screening program. VHWs screened 1335 children in Group A and 1480 children in Group B. The refer rate for 2nd screening was very low (0.8%); the follow-up rate for 2nd screening was between 80 and 97% across the different age groups. Integration of tele-ABR resulted in 11% improvement in follow-up compared to in-person ABR at a tertiary care hospital.
CONCLUSIONS: Non-availability of audiologists and limited infrastructure in rural areas has prevented the establishment of large scale hearing screening programs. In existing programs, considerable challenges with respect to follow-up for diagnostic testing was reported, due to patients being submitted to traveling long distance to access services and potential wage losses during that time. In this program model, integration of a tele-ABR diagnostic follow-up improved follow-up in comparison to in-person follow-up. VHWs were successfully trained to conduct accurate screenings in rural communities. The very low refer rate, and improved follow-up rate reflect the success of this community-based hearing screening program.

Rashid SMU, Mukherjee D, Ahmmed AU.
Auditory processing and neuropsychological profiles of children with functional hearing loss.
Int J Pediatr Otorhinolaryngol. 2018 Nov;114:51-60. doi: 10.1016/j.ijporl.2018.07.054. Epub 2018 Jul 31.

OBJECTIVES: This paper compares structured history, auditory processing abilities and neuropsychological findings of children with functional hearing loss (FHL) to those with suspected auditory processing disorder without FHL (control). The main aim was to evaluate the value of a holistic assessment protocol for FHL used in

a routine pediatric audiology clinic. The protocol incorporated a commercially available test battery for auditory processing disorder (APD), non-verbal intelligence (NVIQ) and tools to screen for common co-existing neurodevelopmental conditions such as attention deficit hyperactivity disorder (ADHD), language impairment (LI) and developmental coordination disorder (DCD). The outcome of such holistic assessment was expected to help in understanding the nature of FHL and to provide individualized support to mitigate their difficulties.
METHODS: This retrospective study compared two groups, 40 children (M = 17, F = 23) in each group between seven and sixteen years of age, one group with a history of FHL and the other with suspected APD without FHL (control). The groups were matched against age, gender, hand use, diagnosis of APD or non-APD (31 with APD and 9 without APD in each group) and non-verbal intelligence. All the children were healthy English speaking children attending mainstream schools with no middle or inner ear abnormalities. Structured history was obtained from parents regarding different nonacademic and academic concerns. The SCAN-3:C and SCAN-3:A test batteries were used to assess auditory processing abilities; Lucid Ability test for NVIQ; Children's Communication Checklist-2 (CCC-2) for language ability; Swanson Nolan and Pelham-IV Rating Scale (SNAP-IV) for ADHD; and the manual dexterity components of the Movement Assessment Battery for Children-2 (MABC-2) as a screening tool for DCD.
RESULTS: About 60% of children in both the groups had concerns regarding listening in noisy background. In the history, poor attention was reported in 45% of children in the FHL group compared to 82.5% in the control group ($p < 0.01$). Hyperacusis was present in 35% of children in the FHL group and in 62% of children in the control group ($p < 0.05$). Concerns about overall academic abilities were present in 59% of children in the FHL group and 75% of the controls ($p > 0.05$). Only 15% of children in the FHL group had concerns with numeracy skills in contrast to 41% of the controls ($p < 0.05$). Significantly fewer ($p < 0.01$) children in the FHL group (41%) received additional support at school than the controls (75%). Fewer children performed poorly in Filtered Words (FW) test of the SCAN-3 batteries, 30% in the FHL group and 17.5% in the control group, in contrast to Auditory Figure Ground 0 (AFG0), 85% in FHL and 80% in the control group. The number of children performing poorly in AFG0 was significantly higher compared to all the other SCAN-3 tests in FHL ($P < 0.05$), in contrast to FW and Competing Sentences (CS) only in the control group ($p < 0.05$). The control group had higher prevalence of atypical ear advantage (AEA) in left directed Competing Words (CW) (32.5%) and Time Compressed Sentences (TCS) (32.5%) compared to FW (7.5%). In contrast, FHL group had higher prevalence of AEA in AFG0 (48.7%) compared to CS (21%). High proportions of children in both the groups had LI (80% in FHL and 82.5% in the control group), with significantly lower ($p < 0.05$) levels of ADHD symptoms in the FHL group (39.5%) compared to the control group (72.5%). Impaired manual dexterity was present in 30.7% of children in FHL group and 47.5% in the controls.
CONCLUSIONS: The prevalences of APD and language impairment are high compared to ADHD symptoms in children with FHL, and holistic assessment is recommended. Despite some similarities in the auditory and neuropsychological profiles between children with FHL and those with suspected APD without FHL some differences were noted. The results suggest that children with FHL have genuine difficulties that need to be identified and addressed. Future research is required to identify the neural pathways which could explain the similarities and dissimilarities between the two groups.

Reis FMFDS, Gonçalves CGO, Conto J, Iantas M, Lüders D, Marques J.
Hearing Assessment of Neonates at Risk for Hearing Loss at a Hearing Health High Complexity Service: An Electrophysiological Assessment.
Int Arch Otorhinolaryngol. 2019 Apr;23(2):157-164. doi: 10.1055/s-0038-1648217. Epub 2018 May 9.
INTRODUCTION: Hearing is the main sensory access in the first years of life. Therefore, early detection and intervention of hearing impairment must begin before the first year of age.
OBJECTIVE: To analyze the results of the electrophysiological hearing assessment of children at risk for hearing loss as part of the newborn hearing screening (NHS).
METHODS: This is a cross-sectional study held at a hearing health public service clinic located in Brazil, with 104 babies at risks factors for hearing loss referred by public hospitals. A questionnaire was applied to parents, and the auditory brainstem response (ABR) test was held, identifying those with alterations in the results. The outcome of the NHS was also analyzed regarding risk factor, gestational age and gender.
RESULTS: Among the 104 subjects, most of them were male (53.85%), and the main risk factor found was the admission to the neonatal intensive care unit (NICU) for a period longer than 5 days (50.93%). Eighty-five (81.73%) subjects were screened by NHS at the maternity and 40% of them failed the test. Through the ABR test, 6 (5.77%) infants evidenced sensorineural hearing loss, 4 of them being diagnosed at 4 months, and 2 at 6 months of age; all of them failed the NHS and had family history and admission at NICU for over 5 days as the most prevalent hearing risks; in addition, family members of all children perceived their hearing impairment.
CONCLUSION: Advances could be observed regarding the age of the diagnosis after the implementation of the NHS held at the analyzed public service clinic.

Riga M, Korres G, Chouridis P, Naxakis S, Danielides V.
Congenital cytomegalovirus infection inducing non-congenital sensorineural hearing loss during childhood; A systematic review.
Int J Pediatr Otorhinolaryngol. 2018 Dec;115:156-164. doi: 10.1016/j.ijporl.2018.10.005. Epub 2018 Oct 4.

BACKGROUND: Congenital cytomegalovirus (CMV) infection is one of the most important risk factors for delayed onset and progressive hearing loss in children. However, the relevant literature is limited, heterogeneous and currently insufficient to provide guidance toward the effective monitoring of hearing acuity in these children.
OBJECTIVES: The aim of this study was to provide a systematic review focused on types of hearing loss that may escape diagnosis through universal neonatal hearing screening and/or present significant changes during childhood, such as progressive, fluctuating and late-onset hearing loss.
DATA SOURCES: A review of the present literature was conducted via the PubMed database of the US National Library of Medicine (www.pubmed.org) and Scopus database (www.scopus.com) with the search terms "late-onset hearing loss cytomegalovirus", "progressive hearing loss cytomegalovirus" and "fluctuating hearing loss cytomegalovirus".
STUDY ELIGIBILITY CRITERIA: Prospective or retrospective clinical studies were included if they presented a detailed audiological assessment, for a follow-up period of >2years.
METHODS: The prevalence and time of diagnosis of progressive, fluctuating and late-onset hearing loss were considered as primary outcomes. Results were recorded separately for symptomatic and asymptomatic children, when possible.
RESULTS: This analysis refers to a population of 181 children with CMV-induced hearing loss, who were diagnosed among 1089 with congenital CMV infection. The prevalence of CMV-induced hearing loss was significantly higher among symptomatic children ($p < 0.0001$), who were also significantly more likely to develop bilateral hearing loss ($p = 0.001$). There was not sufficient information on the prevalence, laterality, degree and time of diagnosis of progressive, fluctuating and late-onset hearing loss that could constitute the basis toward the report of specific follow-up guidelines.
CONCLUSIONS: Further studies are needed in order to understand and quantify the potential effects of congenital CMV infection in the inner ear and hearing acuity. The results presented in the relative studies should be very carefully evaluated and compared to each other, since they correspond to substantially different cohorts, study designs, and result elaboration. Infants with congenital CMV infection should be closely monitored, regarding their hearing acuity at least during their preschool years, although substantial changes in hearing thresholds have been reported as late as the 16th year of age. Parental counseling is of outmost importance in order to minimize the numbers of children lost to follow-up.

Rissatto-Lago MR, da Cruz Fernandes L, Lyra IM, Terse-Ramos R, Teixeira R, Salles C, Teixeira Ladeia AM.
Hidden hearing loss in children and adolescents with sickle cell anemia.
Int J Pediatr Otorhinolaryngol. 2019 Jan;116:186-191. doi: 10.1016/j.ijporl.2018.10.042. Epub 2018 Nov 2.

OBJECTIVE: To evaluate the auditory system for hidden hearing loss (HHL) and its association with clinical variables and endothelial dysfunction (ED) in children and adolescents with sickle cell anemia (SCA).
METHODS: Participants included 37 patients with stable SCA and 44 healthy controls (HC group) (aged 6-18 years) with hearing thresholds ≤ 20 dB (dB) were evaluated for pure tone audiometry, tympanometry, acoustic reflex, otoacoustic emission, and auditory evoked potentials. Laboratory analysis of the lipid profile, and C-reactive protein levels and endothelial function using ultrasonographic imaging of the brachial artery to assess flow-mediated dilation were performed.
RESULTS: The SCA group presented with a higher rate of increased contralateral acoustic reflex thresholds, compared to those in the HC group at all frequencies and in both ears ($p < 0.05$). There were significant differences in the brainstem auditory evoked potentials between the SCA and HC groups. In the SCA group, the waves III and V latencies were increased ($p = 0.006$ and 0.004 respectively), and the I-III and I-V interpeak intervals were longer ($p = 0.015$ and 0.018 respectively) than those in the HC group. There was no association between the audiological measures and clinical and metabolic variables and sickle cell anemia complications including endothelial function and therapy.
CONCLUSION: In conclusion, our findings suggest that damage in the auditory system in SCA patients can be present involving retrocochlear structures, causing functional deficits without deterioration of auditory sensitivity.

Robertson MS, Hayashi SS, Camet ML, Trinkaus K, Henry J, Hayashi RJ.
Asymmetric sensorineural hearing loss is a risk factor for late-onset hearing loss in pediatric cancer survivors following cisplatin treatment.
Pediatr Blood Cancer. 2019 Jan;66(1):e27494. doi: 10.1002/pbc.27494. Epub 2018 Oct 18.

BACKGROUND: Ototoxicity is a significant complication of cisplatin treatment. Hearing loss can be symmetric or asymmetric, and may decline after therapy. This study examined the risks of asymmetric and late-onset hearing loss (LOHL) in cisplatin-treated pediatric patients with cancer.

METHODS: A retrospective review of 993 patients' medical and audiological charts from August 1990 to March 2015 was conducted using stringent criteria to characterize patients with asymmetric hearing loss (AHL) or LOHL. Audiologic data were reviewed for 248 patients that received cisplatin to assess cisplatin-induced sensorineural hearing loss and its associated risk factors.
RESULTS: Of the patients evaluable for AHL, 26% exhibited this finding. Of those evaluable for LOHL, 42% of the patients' hearing worsened more than 6 months after therapy completion. Radiation and type of cancer diagnosis were major risk factors for both AHL and LOHL. Furthermore, LOHL was linked to age of diagnosis, noncranial radiation, and longer audiologic follow-up. AHL was strongly associated with LOHL-60% of patients with AHL also had LOHL. Logistic regression analysis revealed that patients with AHL (OR 6.3, 95% CI: 2.2-17.8, P = 0.0005) or those receiving radiation (OR 3.2, 95% CI: 1.2-8.6, P = 0.02) were at greatest risk for LOHL.
CONCLUSION: Children receiving cisplatin therapy are at risk for developing AHL and LOHL. Those that have received radiation and/or with AHL are at increased risk for further hearing decline. Long-term monitoring of these patients is important for early intervention as hearing diminishes.

Ropers FG, Pham ENB, Kant SG, Rotteveel LJC, Rings EHHM, Verbist BM, Dekkers OM.
Assessment of the Clinical Benefit of Imaging in Children With Unilateral Sensorineural Hearing Loss: A Systematic Review and Meta-analysis.
JAMA Otolaryngol Head Neck Surg. 2019 Apr 4. doi: 10.1001/jamaoto.2019.0121. [Epub ahead of print]

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.

Schaefer K, Coninx F, Fischbach T.
LittlEARS auditory questionnaire as an infant hearing screening in Germany after the newborn hearing screening.
Int J Audiol. 2019 Apr 23:1-8. doi: 10.1080/14992027.2019.1597287. [Epub ahead of print]

OBJECTIVE: To investigate the feasibility of using the LittlEARS® Auditory Questionnaire (LEAQ®) as part of the infant hearing screening programme in Germany.
DESIGN: LEAQ®s were distributed to 47 paediatric practices and were completed by the parents/guardians of the infants (aged between 9-14 months) involved in the study (= LEAQ® screening). The infants who failed the LEAQ® screening were invited to a LEAQ rescreening. Infants who failed the LEAQ® rescreening were sent to a paediatric ENT specialist. After 3 years, a follow-up was performed on two groups: the first group comprised infants who failed the LEAQ screening; the second group (control group) comprised 200 infants who passed the LEAQ screening.
STUDY SAMPLE: 5316 questionnaires were returned.
RESULTS: Six infants with permanent hearing loss were identified using the LEAQ® as a screening tool.

CONCLUSIONS: An infant hearing screening using the LEAQ® is easily implementable in paediatric practices and may be a good alternative in countries where no objective screening instruments are available. The LEAQ® was suitable for monitoring hearing development in infants in general and could help to identify a late-onset or progressive hearing loss in infants.

Schopper HK, D'Esposito CF, Muus JS, Kanter J, Meyer TA.
Childhood Hearing Loss in Patients With Sickle Cell Disease in the United States.
J Pediatr Hematol Oncol. 2019 Mar;41(2):124-128. doi: 10.1097/MPH.0000000000001373.

ABSTRACT: This study sought to examine if modern medical evaluations including newborn screening and early diagnosis along with better methods of disease control have improved rates of hearing loss in children with sickle cell disease (SCD). Audiometric and medical data for patients with SCD was obtained from the AudGen Database and analyzed for the presence of hearing loss, type of hearing loss, severity of hearing loss, and correlation with comorbid conditions. Children with sickle cell trait (SCT) were used as a comparison group. A total of 189 patients with SCD and 244 patients with SCT had sufficient audiologic data available. Hearing loss was present in 62% of children with SCD and 50% of children with SCT in the study population. Patients with SCD were significantly more likely than those with SCT to have a sensorineural component to their hearing loss ($P<0.001$, odds ratio: 2.41 [1.53 to 3.79]) and to have severe or profound hearing loss ($P=0.02$, odds ratio: 4.00 [1.14 to 14.04]). The true prevalence of hearing loss in children with SCD has not been established as routine screening is not being performed. Routine auditory testing should be done for these children to detect this loss before it impacts development.

Shah J, Pham GN, Zhang J, Pakanati K, Raol N, Ongkasuwan J, Hopkins B, Anne S.
Evaluating diagnostic yield of computed tomography (CT) and magnetic resonance imaging (MRI) in pediatric unilateral sensorineural hearing loss.
Int J Pediatr Otorhinolaryngol. 2018 Dec;115:41-44. doi: 10.1016/j.ijporl.2018.09.003. Epub 2018 Sep 11.

INTRODUCTION: Options for imaging for evaluation of pediatric patients with unilateral sensorineural hearing loss (USNHL) include computed tomography (CT) and magnetic resonance imaging (MRI). Although both CT and MR imaging provide valuable information in the evaluation of pediatric patients with USNHL, debate remains regarding which imaging modality is most ideal and should be the preferred study for these children. The objective of this study is to evaluate and compare the diagnostic yield of CT versus MRI in children with USNHL.
METHODS: A multi-institutional retrospective chart review was conducted. Pediatric patients with hearing loss (diagnosis codes 389.00-389.22) seen between 2010 and 2012 at three tertiary care centers were identified. Only patients with USNHL and imaging studies were reviewed and results of CT and MRI for each patient were examined and compared. Cochleovestibular or central nervous system findings known to directly correlate to SNHL were noted as positive findings on imaging. McNemar's test was used to compare patients with positive CT and MRI results.
RESULTS: A total of 219 patients between the ages of 0-18 years with USNHL who underwent CT and/or MRI were identified. Imaging abnormalities were found in 41/96 patients who underwent MRI with overall diagnostic yield of 42.7% and 69 of 188 patients who underwent CT with overall diagnostic yield of 36.7%. For patients who underwent both imaging modalities ($n = 65$), there was no statistically significant difference in positive findings detected by CT vs MRI ($p > 0.05$).
CONCLUSIONS: Both CT and MR imaging have similar overall diagnostic yield when used to evaluate children with USNHL. Parents and patients should be counseled regarding cost, test duration, radiation exposure, need for sedation, and diagnostic accuracy associated with each imaging modality and these factors should be considered to select the appropriate diagnostic study.

Sharma R, Gu Y, Ching TYC, Marnane V, Parkinson B.
Economic Evaluations of Childhood Hearing Loss Screening Programmes: A Systematic Review and Critique.
Appl Health Econ Health Policy. 2019 Jan 25. doi: 10.1007/s40258-018-00456-1. [Epub ahead of print]

BACKGROUND: Permanent childhood hearing loss is one of the most common birth conditions associated with speech and language delay. A hearing screening can result in early detection and intervention for hearing loss.
OBJECTIVES: To update and expand previous systematic reviews of economic evaluations of childhood hearing screening strategies, and explore the methodological differences.
DATA SOURCES: MEDLINE, Embase, the Cochrane database, National Health Services Economic Evaluation Database (NHS EED), the Health Technology Assessment (HTA) database, and Canadian Agency for Drugs and Technologies in Health's (CADTH) Grey matters.
STUDY ELIGIBILITY CRITERIA, PARTICIPANTS AND INTERVENTIONS: Economic evaluations reporting costs and outcomes for both the intervention and comparator arms related to childhood hearing screening strategies.
RESULTS: Thirty evaluations (from 29 articles) were included for review. Several methodological issues were identified, including: few evaluations reported outcomes in terms of quality-adjusted life years (QALYs); none

estimated utilities directly from surveying children; none included disutilities and costs associated with adverse events; few included costs and outcomes that differed by severity; few included long-term estimates; none considered acquired hearing loss; some did not present incremental results; and few conducted comprehensive univariate or probabilistic sensitivity analysis. Evaluations published post-2011 were more likely to report QALYs and disability-adjusted life years (DALYs) as outcome measures, include long-term treatment and productivity costs, and present incremental results.
LIMITATIONS: We were unable to access the economic models and, although we employed an extensive search strategy, potentially not all relevant economic evaluations were identified.
CONCLUSIONS AND IMPLICATIONS: Most economic evaluations concluded that childhood hearing screening is value for money. However, there were significant methodological limitations with the evaluations.

Shekdar KV, Bilaniuk LT.
Imaging of Pediatric Hearing Loss.
Neuroimaging Clin N Am. 2019 Feb;29(1):103-115. doi: 10.1016/j.nic.2018.09.011. Epub 2018 Oct 31.
ABSTRACT: Temporal bone high-resolution computed tomography (HRCT) and magnetic resonance (MR) imaging are valuable tools in the evaluation of pediatric hearing loss. Computed tomography is important in the evaluation of pediatric conductive hearing loss and is the imaging modality of choice for evaluation of osseous abnormalities. MR imaging is the modality of choice for evaluation of sensorineural hearing loss. A broad spectrum of imaging findings can be seen with hearing loss in children. HRCT and MR imaging provide complementary information and are often used in conjunction in the preoperative evaluation of pediatric candidates for cochlear implantation.

Sheppard S, Biswas S, Li MH, Jayaraman V, Slack I, Romasko EJ, Sasson A, Brunton J, Rajagopalan R, Sarmady M, Abrudan JL, Jairam S, DeChene ET, Ying X, Choi J, Wilkens A, Raible SE, Scarano MI, Santani A, Pennington JW, Luo M, Conlin LK, Devkota B, Dulik MC, Spinner NB, Krantz ID.
Utility and limitations of exome sequencing as a genetic diagnostic tool for children with hearing loss.
Genet Med. 2018 Dec;20(12):1663-1676. doi: 10.1038/s41436-018-0004-x. Epub 2018 Jun 15.
PURPOSE: Hearing loss (HL) is the most common sensory disorder in children. Prompt molecular diagnosis may guide screening and management, especially in syndromic cases when HL is the single presenting feature. Exome sequencing (ES) is an appealing diagnostic tool for HL as the genetic causes are highly heterogeneous.
METHODS: ES was performed on a prospective cohort of 43 probands with HL. Sequence data were analyzed for primary and secondary findings. Capture and coverage analysis was performed for genes and variants associated with HL.
RESULTS: The diagnostic rate using ES was 37.2%, compared with 15.8% for the clinical HL panel. Secondary findings were discovered in three patients. For 247 genes associated with HL, 94.7% of the exons were targeted for capture and 81.7% of these exons were covered at 20× or greater. Further analysis of 454 randomly selected HL-associated variants showed that 89% were targeted for capture and 75% were covered at a read depth of at least 20×.
CONCLUSION: ES has an improved yield compared with clinical testing and may capture diagnoses not initially considered due to subtle clinical phenotypes. Technical challenges were identified, including inadequate capture and coverage of HL genes. Additional considerations of ES include secondary findings, cost, and turnaround time.

Siu JM, Blaser SI, Gordon KA, Papsin BC, Cushing SL.
Efficacy of a selective imaging paradigm prior to pediatric cochlear implantation.
Laryngoscope. 2019 Jan 6. doi: 10.1002/lary.27666. [Epub ahead of print]
OBJECTIVES/HYPOTHESIS: There is no consensus on the necessary preoperative imaging in children being evaluated for cochlear implantation (CI). Dual-imaging protocols that implement both magnetic resonance imaging (MRI) and high resolution computed tomography (HRCT) create diagnostic redundancy in the face of potentially unnecessary radiation and anaesthetic exposure. The objectives of the current study were to examine the efficacy of an MRI-predominant with selective HRCT imaging protocol.
STUDY DESIGN: Retrospective review.
METHODS: The protocol was implemented over a 4-year period, during which HRCT was obtained in addition to MRI only if specific risk factors on clinical assessment were identified or if imaging findings in need of further evaluation were detected on initial MRI evaluation. Retrospective review of operative reports and prospective review of imaging were performed; anesthetic exposure and costing information were also obtained.
RESULTS: Of the 240 patients who underwent assessment, seven (2.9%) had combined HRCT and MRI performed concurrently based on initial clinical assessment, 15 (6.3%) underwent HRCT based on imaging anomalies found on MRI, and MRI alone was ordered for the remaining 218 (90.1%). All patients were implanted without complication. Overall, radiation exposure, general anesthesia (GA), and healthcare costs were reduced.

CONCLUSIONS: MRI alone can be used in the vast majority of cases for preoperative evaluation of pediatric CI candidates resulting in a significant reduction in healthcare costs, radiation, and GA exposure in children. The additional need for HRCT occurs in a small proportion and can be predicted up front on clinical assessment or on initial MRI.

Skou AS, Olsen SØ, Nielsen LH, Glosli H, Jahnukainen K, Jarfelt M, Jónmundsson GK, Malmros J, Nysom K, Hasle H; Nordic Society of Pediatric Hematology and Oncology (NOPHO).
Hearing Status in Survivors of Childhood Acute Myeloid Leukemia Treated With Chemotherapy Only: A NOPHO-AML Study.
J Pediatr Hematol Oncol. 2019 Jan;41(1):e12-e17. doi: 10.1097/MPH.0000000000001302.

BACKGROUND: As more children survive acute myeloid leukemia (AML) it is increasingly important to assess possible late effects of the intensive treatment. Hearing loss has only sporadically been reported in survivors of childhood AML. We assessed hearing status in survivors of childhood AML treated with chemotherapy alone according to 3 consecutive NOPHO-AML trials.
PROCEDURE: A population-based cohort of children treated according to the NOPHO-AML-84, NOPHO-AML-88, and NOPHO-AML-93 trials included 137 eligible survivors among whom 101 (74%) completed a questionnaire and 99 (72%) had otologic and audiologic examination performed including otoscopy (72%), pure tone audiometry (70%), and tympanometry (60%). Eighty-four of 93 (90%) eligible sibling controls completed a similar questionnaire.
RESULTS: At a median of 11 years (range, 4 to 25) after diagnosis, hearing disorders were rare in survivors of childhood AML and in sibling controls, with no significant differences. None had severe or profound hearing loss diagnosed at audiometry. Audiometry detected a subclinical hearing loss ranging from slight to moderate in 19% of the survivors, 5% had low-frequency hearing loss, and 17% had high-frequency hearing loss.
CONCLUSIONS: The frequency of hearing disorders was low, and hearing thresholds in survivors of childhood AML were similar to background populations of comparable age.

Sokolov M, Gordon KA, Polonenko M, Blaser SI, Papsin BC, Cushing SL.
Vestibular and balance function is often impaired in children with profound unilateral sensorineural hearing loss.
Hear Res. 2019 Feb;372:52-61. doi: 10.1016/j.heares.2018.03.032. Epub 2018 Apr 3.

RATIONALE: Children with unilateral deafness could have concurrent vestibular dysfunction which would be associated with balance deficits and potentially impair overall development. The prevalence of vestibular and balance deficits remains to be defined in these children.
METHODS: Twenty children with unilateral deafness underwent comprehensive vestibular and balance evaluation.
RESULTS: Retrospective review revealed that more than half of the cohort demonstrated some abnormality of the vestibular end organs (otoliths and horizontal canal), with the prevalence of end organ specific dysfunction ranging from 17 to 48% depending on organ tested and method used. In most children, impairment occurred only on the deaf side. Children with unilateral deafness also displayed significantly poorer balance function than their normal hearing peers.
CONCLUSIONS: The prevalence of vestibular dysfunction in children with unilateral deafness is high and similar to that of children with bilateral deafness. Vestibular and balance evaluation should be routine and the functional impact of combined vestibulo-cochlear sensory deficits considered.

Soylemez E, Ertugrul S, Dogan E.
Assessment of balance skills and falling risk in children with congenital bilateral profound sensorineural hearing loss.
Int J Pediatr Otorhinolaryngol. 2019 Jan;116:75-78. doi: 10.1016/j.ijporl.2018.10.034. Epub 2018 Oct 23.

OBJECTIVE: To evaluate the balance skills and falling risk in children with a congenital bilateral profound sensorineural hearing loss (CBPSNHL).
METHODS: 25 children with CBPSNHL and healthy 25 children with similar age and gender were included in the study. The flamingo balance test, the tandem stance test, and the one-leg standing test were performed to assess the patients' static balance skills. The pediatric balance scale (PBS) was used to evaluate the dynamic balance. Visual analog scale (VAS) was applied to the patients assess the frequency of falls.
RESULTS: The flamingo balance test, the tandem stance test, and the one-leg standing test in the children with CBPSNHL were all significantly worse than the control group. Although the scores of PBS in patients with CBPSNHL were significantly lower than the control group ($p < 0.001$), the results of both groups were consistent with a low risk of falls. There was no significant difference between the VAS scores indicating the frequency of falls among the groups ($p = 0.552$).
CONCLUSION: Static and dynamic balance skills of the children with CBPSNHL are significantly impaired compared to their healthy peers. Children with CBPSNHL also have a lower risk of falling just like their healthy

peers and there is no significant difference between their falling frequencies. Balance skills of children with CBPSNHL can be assessed quickly and effectively on a hard floor (eyes closed), with a tandem standing test or a one-leg standing test.

Sözen T, Bajin MD1, Kara A, Sennaroğlu L.
The Effect of National Pneumococcal Vaccination Program on Incidence of Postmeningitis Sensorineural Hearing Loss and Current Treatment Modalities.
J Int Adv Otol. 2018 Dec;14(3):443-446. doi: 10.5152/iao.2018.6169.

OBJECTIVES: The aim of the present study was to investigate the effect of the national pneumococcal vaccination program on postmeningitis sensorineural hearing loss (SNHL).
MATERIALS AND METHODS: Overall, 2751 patients (2615 cochlear implantation and 136 auditory brainstem implantation) who underwent cochlear implantation (CI) and auditory brainstem implantation (ABI) at a tertiary referral hospital otolaryngology clinic were retrospectively analyzed. One hundred sixteen patients with a history of meningitis were included in the study. Patients were evaluated for their age at the time of surgery, gender, computerized tomography (CT) and magnetic resonance imaging (MRI) findings, implant type, side, and incidence before and after the vaccination program.
RESULTS: When patients with cochlear implants or ABI were examined, the incidence of meningitis-induced hearing loss was 6.2% in the pre-vaccination period and 0.6% in the post-vaccination period. There is a significant difference between them when compared by chi-square test ($p<0.001$).
CONCLUSION: The most important finding of the present study is the dramatic decrease in the number of CI and ABI surgeries performed in patients with SNHL due to meningitis. This shows the effectivity of pneumococcal vaccination in this special group of patients. If total ossification is detected on CT of patients with postmeningitis, ABI should be preferred to CI.

Steuerwald W, Windmill I, Scott M, Evans T, Kramer K.
Stories From the Webcams: Cincinnati Children's Hospital Medical Center Audiology Telehealth and Pediatric Auditory Device Services.
Am J Audiol. 2018 Nov 19;27(3S):391-402. doi: 10.1044/2018_AJA-IMIA3-18-0010.

PURPOSE: The purpose of this manuscript is to describe the regulatory, technological, and training considerations for audiologists investigating telehealth and to offer some examples of audiology services provided through telehealth.
METHOD: The authors presented the regulatory components, the technology required for audiology staff and patients, and staff training for the audiology telehealth program at Cincinnati Children's Hospital Medical Center. Four case studies highlighting the successful use of telehealth in providing auditory device services to patients were also presented.
RESULTS AND CONCLUSION: The described regulatory, technological, and training hierarchy provides a framework for audiologists interested in starting a telehealth program. The cases presented illustrate that telehealth can be used to provide some auditory device services, such as troubleshooting, mapping, and parent consulting.

Stewart JE, Bentley JE.
Hearing Loss in Pediatrics: What the Medical Home Needs to Know.
Pediatr Clin North Am. 2019 Apr;66(2):425-436. doi: 10.1016/j.pcl.2018.12.010.

ABSTRACT: Screening infants for hearing loss at birth is a standard in most states in the United States, but follow-up continues to warrant improvement. Understanding the definition of hearing loss, its etiology, appropriate intervention options, and knowledge of methods to optimize an infant's outcomes through the medical home can help to maximize speech and language skills.

Tang K, Gao Z, Han C, Zhao S, Du X, Wang W.
Screening of mitochondrial tRNA mutations in 300 infants with hearing loss.
Mitochondrial DNA A DNA Mapp Seq Anal. 2019 Mar;30(2):345-350. doi: 10.1080/24701394.2018.1527910. Epub 2018 Nov 19.

ABSTRACT: Mitochondrial DNA (MtDNA) mutations are the important causes for hearing loss. To see the contribution of mtDNA to deafness, we screened for mutations in mt-tRNA genes from 300 deaf infants and 200 healthy subjects. Moreover, we analyzed the mtDNA copy number and ROS levels in patients carrying the mt-tRNA mutations. Consequently, 3 mt-tRNA mutations: $tRNA^{Leu(UUR)}$ A3243G; $tRNA^{Ala}$ T5655C and $tRNA^{Glu}$ A14692G were identified, however, these mutations were not detected in controls. Of these, the A3243G mutation created a novel base-pairing (13G-23A) in the D-stem of $tRNA^{Leu(UUR)}$; while the T5655C mutation occurred at the very conserved acceptor arm of $tRNA^{Ala}$; in addition, the A14692G mutation was located at position 55 in the TΨC loop of $tRNA^{Glu}$. Molecular analysis showed that patients harbouring the A3243G, T5655C

and A14692G mutations had a lower level of mtDNA copy number, while ROS level increased significantly when compared with controls. Through the application of the pathogenicity scoring system, we noticed that the A3243G, T5655C and A14692G should be regarded as 'definitely pathogenic' mutations associated with deafness. Thus, our study provided novel insight into the pathophysiology, early detection of mitochondrial deafness.

Towerman AS, Hayashi SS, Hayashi RJ, Hulbert ML.
Prevalence and nature of hearing loss in a cohort of children with sickle cell disease.
Pediatr Blood Cancer. 2019 Jan;66(1):e27457. doi: 10.1002/pbc.27457. Epub 2018 Sep 11.

BACKGROUND: Sickle cell disease (SCD) may cause injury to any organ, including the auditory system. Although the association of SCD and hearing loss has been described, the nature of this complication is unknown. We sought to establish the prevalence and nature of hearing loss in a referred cohort of children with SCD and to identify correlating disease- or treatment-associated factors.
PROCEDURE: We conducted a retrospective review of patients with SCD < 22 years of age who had hearing evaluations between August 1990 and December 2014. Demographics, audiograms, and disease and treatment variables were analyzed.
RESULTS: Two hundred and ten audiograms among 81 patients were reviewed, and 189 were evaluable. Seventy-two children constituted the referred cohort. Fourteen (19.4%) had hearing loss documented on at least one audiogram. Seven (9.7%) patients had only conductive hearing loss, and the loss persisted for up to 10.3 years. The median age of first identification was eight years. Six (8.3%) patients had hearing loss that was at least partially sensorineural. One patient's hearing loss was ambiguous. All sensorineural hearing losses were unilateral and 4/6 patients had prior documented normal hearing, indicating acquired loss. No correlations were identified.
CONCLUSIONS: Both conductive and sensorineural hearing losses are more prevalent in our study population than those observed in the general pediatric population. In children with SCD, sensorineural hearing loss appears to be acquired and unilateral. Conductive hearing loss was identified in older children and can persist. Serial screening is needed for early detection and more prompt intervention in this population.

Tsai YT, Fang KH, Yang YH, Lin MH, Chen PC, Tsai MS, Hsu CM.
Risk of developing sudden sensorineural hearing loss in patients with hepatitis B virus infection: A population-based study.
Ear Nose Throat J. 2018 Oct-Nov;97(10-11):E19-E27.

ABSTRACT: Sudden sensorineural hearing loss (SSNHL) has significant impact on quality of life. It may result from viral infection, but the relationship between hepatitis B virus (HBV) infection and SSNHL remains uncertain. To investigate the risk of developing SSNHL in patients with HBV, we conducted a nationwide, population-based, retrospective cohort study from the Taiwan National Health Insurance Research Database. A total of 33,234 patients diagnosed with HBV infection and 132,936 control subjects without viral hepatitis were selected from claims made from 2000 to 2008. Each patient was followed for at least 5 years to identify new-onset SSNHL. Among the 166,170 patients, 279 patients (303,793 person-years) from the HBV cohort and 845 patients (1,225,622 person-years) from the control cohort were diagnosed with SSNHL. The incidence of SSNHL was 1.33-fold higher in the HBV group than in the control group (0.92 vs. 0.69 per 10,000 person-years), with an adjusted hazard ratio (HR) of 1.315 (95% confidence interval [CI] = 1.148 to 1.506) calculated using a Cox proportional hazard regression model. We also observed that HBV patients in the 50 to 64 years of age subgroup showed the highest incidence of SSNHL and the highest adjusted hazard ratio (HR = 2.367; 95% CI = 1.958 to 2.861). Patients with HBV infection had a higher risk of acquiring SSNHL than patients without viral hepatitis. For the early detection and timely treatment of SSNHL, clinicians should be aware of the increased risk of SSNHL in HBV patients and arrange auditory examinations for those complaining about acute hearing change.
PMID:

van Hövell Tot Westerflier CVA, van Heteren JAA, Breugem CC, Smit AL, Stegeman I.
Impact of unilateral congenital aural atresia on academic Performance: A systematic review.
Int J Pediatr Otorhinolaryngol. 2018 Nov;114:175-179. doi: 10.1016/j.ijporl.2018.09.002. Epub 2018 Sep 8.

BACKGROUND: Little is known about the academic performance of children with unilateral congenital aural atresia (CAA).
OBJECTIVE: of review: Our objective was to summarize what is known about the academic performance of children with hearing loss by unilateral congenital aural atresia, in order to provide pragmatic recommendations to clinicians who see children with this entity.
TYPE OF REVIEW: Systematic review.
SEARCH STRATEGY: We conducted a systematic search in PubMed Medline, EMBASE, and Cochrane Library combining the terms "atresia" and synonyms with "unilateral hearing loss" and synonyms. Date of the most

recent search was 16 May 2018.
EVALUATION METHOD: Two independent authors identified studies, extracted data, and assessed risk of bias. This review was reported according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA). Observational studies on the academic achievements of patients of any age with unilateral conductive hearing loss of any level due to congenital aural atresia were included. We considered grade retention, special education, individualized education plans, and parental report of school performance as outcome measures for academic achievement.
RESULTS: Two studies reporting on academic performance of patients with unilateral CAA, which both had a significant risk of bias. One study (n = 140) showed a grade retention rate of 3.6% (n = 5) in total. 15.7% (n = 22) needed special education, and 36.4% (n = 51) used an individualized education program. The second study, reporting on 67 patients with unilateral CAA, showed that 29.9% (n = 20) of the patients received school intervention, and 25.4% (n = 17) had learning problems.
CONCLUSION: Current evidence regarding the effect of unilateral congenital aural atresia on academic performance is sparse, inconclusive and has a significant risk of bias. High quality observational studies assessing the effects of aural atresia on academic performance in these patients should be initiated.

Vancor E, Shapiro ED, Loyal J.
Results of a Targeted Screening Program for Congenital Cytomegalovirus Infection in Infants Who Fail Newborn Hearing Screening.
J Pediatric Infect Dis Soc. 2019 Mar 28;8(1):55-59. doi: 10.1093/jpids/pix105.
BACKGROUND: Congenital cytomegalovirus (CMV) infection is a major cause of sensorineural hearing loss. By law, newborns in Connecticut who fail newborn hearing screening are tested for infection with CMV. This targeted screening is controversial, because most children with congenital CMV infection are asymptomatic, and CMV-related hearing loss can have a delayed onset. Our hospital uses a saliva polymerase chain reaction (PCR) assay (confirmed by a urine PCR assay) to detect CMV. Here, we report the results of the first year of our screening program.
METHODS: We reviewed the medical records of newborns in the Yale New Haven Health System who failed the newborn hearing screening test between January 1 and December 31, 2016.
RESULTS: Of 10964 newborns, 171 failed newborn hearing screening, and 3 of these newborns had positive saliva CMV PCR test results. Of these 3 newborns, 2 had positive results on the confirmatory test (for 1 of them the confirmatory test was not performed until the infant was 10 weeks old), and 1 had a negative result on the confirmatory test. Three additional newborns with congenital CMV infection were tested because of clinical indications (1 for ventriculomegaly on prenatal ultrasound and 2 for CMV infection of the mother). Results of audiology follow-up were available for 149 (87.1%) of the 171 newborns who failed newborn hearing screening; 127 (85.2%) had normal results.
CONCLUSION: Our targeted screening program for congenital CMV infection had a low yield. Consideration should be given to other strategies for identifying children at risk of hearing loss as a result of congenital CMV infection.

Vukkadala N, Giridhar SBP, Okumura MJ, Chan DK.
Seeking equilibrium: The experiences of parents of infants and toddlers who are deaf/hard-of-hearing.
J Pediatr Rehabil Med. 2019;12(1):11-20. doi: 10.3233/PRM-170528.
PURPOSE: To identify key determinants of the quality of life of caregivers of infants and toddlers (< 3 years) who are deaf/hard-of-hearing (DHH).
METHODS: We conducted focus groups with providers for children who are DHH as well as interviews with hearing parents of infants and toddlers who are DHH. A multi-step qualitative analysis on interview data using grounded theory was performed, and an iterative analysis to investigate codes to characterize specific topics in caring for deaf infants and toddlers was conducted.
RESULTS: Four focus groups (n= 33) and six semi-structured interviews (n= 7) were conducted. The major theoretical code found was the “Search for Equilibrium” in parenting which arose from the three main categories of the caregiver role/experience: (1) being a parent - modifying parenting style as a result of their child’s hearing loss, (2) being a mediator - modulating and filtering interactions between their child and their child’s environment, and (3) being a navigator - managing the logistics of the medical and educational system.
CONCLUSIONS: For hearing parents, the diagnosis of hearing loss requires changes in multiple domains of parenting. Support in each of these areas is critical for parents to restore a sense of equilibrium that is central to their quality of life. This framework provides a way to categorize parent experiences and may act as a template for focused interventions in the three identified domains.

Walker EA, Curran M, Spratford M, Roush P.
Remote microphone systems for preschool-age children who are hard of hearing: access and utilization.

Int J Audiol. 2019 Apr;58(4):200-207. doi: 10.1080/14992027.2018.1537523. Epub 2019 Jan 5.

OBJECTIVES: Children who are hard of hearing (CHH) have restricted access to auditory-linguistic information. Remote-microphone (RM) systems reduce the negative consequences of limited auditory access. The purpose of this study was to characterise receipt and use of RM systems in young CHH in home and school settings.

DESIGN: Through a combination of parent, teacher, and audiologist report, we identified children who received RM systems for home and/or school use by 4 years of age or younger. With cross-sectional surveys, parents estimated the amount of time the child used RM systems at home and school per day.

STUDY SAMPLE: The participants included 217 CHH.

RESULTS: Thirty-six percent of the children had personal RMs for home use and 50% had RM systems for school. Approximately, half of the parents reported that their children used RM systems for home use for 1-2 hours per use and RM systems for school use for 2-4 hours per day.

CONCLUSIONS: Results indicated that the majority of the CHH in the current study did not receive RM systems for home use in early childhood, but half had access to RM technology in the educational setting. High-quality research studies are needed to determine ways in which RM systems benefit pre-school-age CHH.

Wang LA, Smith PB, Laughon M, Goldberg RN, Ku LC, Zimmerman KO, Balevic S, Clark RH, Benjamin DK, Greenberg RG; Best Pharmaceuticals for Children Act – Pediatric Trials Network Steering Committee.

Prolonged furosemide exposure and risk of abnormal newborn hearing screen in premature infants.

Early Hum Dev. 2018 Oct;125:26-30. doi: 10.1016/j.earlhumdev.2018.08.009. Epub 2018 Sep 4.

BACKGROUND: At very high doses, furosemide is linked to ototoxicity in adults, but little is known about the risk of hearing loss in premature infants exposed to furosemide.

AIMS: Evaluate the association between prolonged furosemide exposure and abnormal hearing screening in premature infants.

STUDY DESIGN: Using propensity scoring, infants with prolonged (≥28 days) exposure to furosemide were matched to infants never exposed. The matched sample was used to estimate the impact of prolonged furosemide exposure on the probability of an abnormal hearing screen prior to hospital discharge.

SUBJECTS: A cohort of infants 501-1250 g birth weight and 23-29 weeks gestational age discharged home from 210 neonatal intensive care units in the United States (2004-2013).

OUTCOME MEASURES: We defined abnormal hearing screen as a result of either “fail” or “refer” for either ear.

RESULTS: Altogether, 1020 infants exposed to furosemide for ≥28 days were matched to 790 unique infants never exposed, yielding a total of 1042 matches due to sampling with replacement and propensity score ties. Matching resulted in a population similar in baseline characteristics. After adjusting for covariates, the proportion of infants with an abnormal hearing screen in the furosemide-exposed group was not significantly higher than the never-exposed group (absolute difference 3.0% [95% CI -0.2-6.2%], P = 0.07).

CONCLUSIONS: Prolonged furosemide exposure was associated with a positive, but not statistically significant, difference in abnormal hearing screening in premature infants. Additional studies with post-hospital discharge audiology follow-up are needed to further evaluate the safety of furosemide in this population.

Wang Q, Xiang J, Sun J, Yang Y, Guan J, Wang D, Song C, Guo L, Wang H, Chen Y, Leng J, Wang X, Zhang J, Han B, Zou J, Yan C, Zhao L, Luo H, Han Y, Yuan W, Zhang H, Wang W, Wang J, Yang H, Xu X, Yin Y, Morton CC, Zhao L1, Zhu S, Shen J, Peng Z.

Nationwide population genetic screening improves outcomes of newborn screening for hearing loss in China.

Genet Med. 2019 Mar 20. doi: 10.1038/s41436-019-0481-6. [Epub ahead of print]

PURPOSE: The benefits of concurrent newborn hearing and genetic screening have not been statistically proven due to limited sample sizes and outcome data. To fill this gap, we analyzed outcomes of newborns with genetic screening results.

METHODS: Newborns in China were screened for 20 hearing-loss-related genetic variants from 2012 to 2017. Genetic results were categorized as positive, at-risk, inconclusive, or negative. Hearing screening results, risk factors, and up-to-date hearing status were followed up via phone interviews.

RESULTS: Following up 12,778 of 1.2 million genetically screened newborns revealed a higher rate of hearing loss by three months of age among referrals from the initial hearing screening (60% vs. 5.0%, P < 0.001) and a lower rate of lost-to-follow-up/documentation (5% vs. 22%, P < 0.001) in the positive group than in the inconclusive group. Importantly, genetic screening detected 13% more hearing-impaired infants than hearing screening alone and identified 2,638 (0.23% of total) newborns predisposed to preventable ototoxicity undetectable by hearing screening.

CONCLUSION: Incorporating genetic screening improves the effectiveness of newborn hearing screening programs by elucidating etiologies, discerning high-risk subgroups for vigilant management, identifying additional children who may benefit from early intervention, and informing at-risk newborns and their maternal relatives of increased susceptibility to ototoxicity.

Wasser J, Ari-Even Roth D, Herzberg O, Lerner-Geva L, Rubin L.
Assessing and monitoring the impact of the national newborn hearing screening program in Israel.
Isr J Health Policy Res. 2019 Mar 11;8(1):30. doi: 10.1186/s13584-019-0296-6.

BACKGROUND: The Israeli Newborn Hearing Screening Program (NHSP) began operating nationally in January 2010. The program includes the Otoacoustic Emissions (OAE) test for all newborns and Automated Auditory Brainstem Response (A-ABR) test for failed OAE and infants at risk for auditory neuropathy spectrum disorders. NHSP targets are diagnosis of hearing impairment by age three months and initiation of habilitation by six months.
OBJECTIVES: (1) Review NHSP coverage; (2) Assess NHSP impact on age at diagnosis for hearing impairment and age at initiation of habilitation; (3) Identify contributing factors and barriers to NHSP success.
METHODS: (1) Analysis of screening coverage and referral rates for the NHSP; (2) Analysis of demographic data, results of coverage, age at diagnosis and initiation of habilitation for hearing impaired infants pre-implementation and post-implementation of NHSP from 10 habilitation centers; (3) Telephone interviews with parents whose infants failed the screening and were referred for further testing.
RESULTS: The NHSP coverage was 98.7% (95.1 to 100%) for approximately 179,000 live births per year for 2014-2016 and average referral rates were under 3%. After three years of program implementation, median age at diagnosis was 3.7 months compared to 9.5 months prior to NHSP. The median age at initiation of habilitation after three years of NHSP was 9.4 months compared to 19.0 prior to NHSP. Parents (84% of 483 sampled) with infants aged 4-6 months participated in the telephone survey. While 84% of parents reported receiving a verbal explanation of the screening results, more than half of the parents reported not receiving written material. Parental report of understanding the test results and a heightened level of concern over the failed screen were associated with timely follow-up.
CONCLUSIONS: The findings indicate high screening coverage. The program reduced ages at diagnosis and initiation of habilitation for hearing impaired infants. Further steps needed to streamline the NHSP are improving communication among caregivers to parents to reduce anxiety; increasing efficiency in transferring information between service providers using advanced technology while ensuring continuum of care; reducing wait time for follow-up testing in order to meet program objectives. Establishment of a routine monitoring system is underway.

Weiss A, Sommer G, Schindera C, Wengenroth L, Karow A, Diezi M, Michel G, Kuehni CE; Swiss Paediatric Oncology Group (SPOG).
Hearing loss and quality of life in survivors of paediatric CNS tumours and other cancers.
Qual Life Res. 2019 Feb;28(2):515-521. doi: 10.1007/s11136-018-2021-2. Epub 2018 Oct 10.

PURPOSE: Hearing loss, a complication of cancer treatment, may reduce health-related quality of life (HRQoL), especially in childhood cancer survivors of central nervous system (CNS) tumours who often have multiple late effects. We examined the effect of hearing loss on HRQoL in young survivors of CNS and other childhood cancers.
METHODS: Within the Swiss Childhood Cancer Survivor Study, we sent questionnaires about hearing loss and HRQoL (KIDSCREEN-27) to parents of survivors aged 8-15 years. We stratified the effect of hearing loss on HRQoL by cancer diagnosis, using multivariable logistic regression and adjusting for sociodemographic and clinical factors.
RESULTS: Hearing loss was associated with impaired physical well-being [unadjusted estimated differences -4.6 (CI -9.2, -0.1); adjusted -4.0 (CI -7.6, -0.3)] and peers and social support [unadjusted -6.7 (CI -13.0, -0.3); adjusted -5.0 (CI -10.5, 0.9)] scores in survivors of CNS tumours (n = 123), but not in children diagnosed with other cancers (all p-values > 0.20, n = 577).
CONCLUSION: Clinicians should be alert to signs of reduced physical well-being and impaired relationships with peers. Especially survivors of CNS tumours may benefit most from strict audiological monitoring and timely intervention to mitigate secondary consequences of hearing loss on HRQoL.

Yazici A, Coskun ME.
The effect of ventilation tube insertion to the health-related quality of life in a group of children in Southeast Anatolia.
Clin Otolaryngol. 2018 Dec;43(6):1578-1582. doi: 10.1111/coa.13220. Epub 2018 Sep 17.

OBJECTIVE: To demonstrate the influence of ventilation tube insertion to the quality of life in a group of children in Southeast Anatolia by Otitis Media 6-item (OM6) questionnaire.
DESIGN: Patients who underwent ventilation tube insertion due to otitis media with effusion (OME) at Otorhinolaryngology Department of Gaziantep University between December 2016 and April 2017 were enrolled in this prospective study. All patients were evaluated with the OM-6 survey before operation and 6 weeks after surgery.
RESULTS: The mean age of 45 patients out of 50 accounted for 67.64 ± 42.89 months with 27 (60%) males and

18 (40%) females. The numbers of preoperative and postoperative overall OM6 scores represented a significant improvement with 4.34 and 2.16, respectively. Moreover, each domain of OM6 (physical suffering, hearing loss, speech impairment, emotional distress, activity limitations and caregiver concerns) showed statistically significant difference.

CONCLUSION: Ventilation tube insertion procedure provided a significant improvement in a group of children in Southeast Anatolia suffering from chronic OME in terms of Quality of Life (QOL) assessed by OM6. We believe that OM6 is a useful tool for evaluating the patients' health-related quality of life and for providing additional information to the caregivers' or families' enquiries regarding the consequences of surgical intervention.

Yimtae K, Israsena P, Thanawirattananit P, Seesutas S, Saibua S, Kasemsiri P, Noymai A, Soonrach T.
A Tablet-Based Mobile Hearing Screening System for Preschoolers: Design and Validation Study.
JMIR Mhealth Uhealth. 2018 Oct 23;6(10):e186. doi: 10.2196/mhealth.9560.

BACKGROUND: Hearing ability is important for children to develop speech and language skills as they grow. After a mandatory newborn hearing screening, group or mass screening of children at later ages, such as at preschool age, is often practiced. For this practice to be effective and accessible in low-resource countries such as Thailand, innovative enabling tools that make use of pervasive mobile and smartphone technology should be considered.

OBJECTIVE: This study aims to develop a cost-effective, tablet-based hearing screening system that can perform a rapid minimal speech recognition level test.

METHODS: An Android-based screening app was developed. The screening protocol involved asking children to choose pictures corresponding to a set of predefined words heard at various sound levels offered in a specifically designed sequence. For the app, the set of words was validated, and their corresponding speech power levels were calibrated. We recruited 122 children, aged 4-5 years, during the development phase. Another 63 children of the same age were screened for their hearing abilities using the app in version 2. The results in terms of the sensitivity and specificity were compared with those measured using the conventional audiometric equipment.

RESULTS: For screening purposes, the sensitivity of the developed screening system version 2 was 76.67% (95% CI 59.07-88.21), and the specificity was 95.83% (95% CI 89.77-98.37) for screening children with mild hearing loss (pure-tone average threshold at 1, 2, and 4 kHz, >20 dB). The time taken for the screening of each child was 150.52 (SD 19.07) seconds (95% CI 145.71-155.32 seconds). The average time used for conventional play audiometry was 11.79 (SD 3.66) minutes (95% CI 10.85-12.71 minutes).

CONCLUSIONS: This study shows the potential use of a tablet-based system for rapid and mobile hearing screening. The system was shown to have good overall sensitivity and specificity. Overall, the idea can be easily adopted for systems based on other languages.

Yoshinaga-Itano C, Sedey AL, Wiggin M, Mason CA.
Language Outcomes Improved Through Early Hearing Detection and Earlier Cochlear Implantation.
Otol Neurotol. 2018 Dec;39(10):1256-1263. doi: 10.1097/MAO.0000000000001976.

HYPOTHESIS: Early identification and intervention, earlier cochlear implantation, and mother's level of education will directly and/or indirectly impact the language outcomes of children with cochlear implants (CIs).

BACKGROUND: Identifying factors that contribute to the wide range of language outcomes in children who use CIs will assist healthcare and rehabilitation professionals in optimizing service delivery for this population. Universal newborn hearing screening provides an opportunity to examine the relationship between meeting the early hearing detection and intervention (EHDI) 1-3-6 guidelines and child language outcomes. These guidelines recommend screening by 1 month, confirmation of hearing loss by 3 months, and intervention by 6 months of age.

METHODS: Participants were 125 children with CIs ranging from 13 to 39 months of age. Language ability was measured using the Child Development Inventory and MacArthur-Bates Communicative Development Inventories.

RESULTS: Meeting EHDI 1-3-6, higher levels of maternal education and earlier cochlear implant activation had a direct, positive impact on language outcomes. Meeting the EHDI 1-3-6 guidelines also had an indirect positive effect on language outcomes via increasing the probability that the children's CIs would be activated earlier. Maternal education did not significantly predict age of cochlear implant activation nor whether a child met EHDI 1-3-6.

CONCLUSION: Ensuring families meet the EHDI 1-3-6 guidelines is an early step that can lead to higher language outcomes and also earlier cochlear implantation.

Zeitler DM, Sladen DP, DeJong MD, Torres JH, Dorman MF, Carlson ML.
Cochlear implantation for single-sided deafness in children and adolescents.
Int J Pediatr Otorhinolaryngol. 2019 Mar;118:128-133. doi: 10.1016/j.ijporl.2018.12.037. Epub 2019 Jan 2.

OBJECTIVE: To evaluate outcomes in pediatric and adolescent patients with single-sided deafness (SSD) undergoing cochlear implantation.
METHODS: A retrospective cohort design at two tertiary level academic cochlear implant centers. The subjects included nine children ages 1.5 to 15 years-old with single-sided deafness (SSD) who had undergone cochlear implantation in the affected ear. Objective outcome measures included were speech reception testing in quiet and noise, bimodal speech reception threshold testing in noise, tinnitus suppression, and device usage.
RESULTS: Nine pediatric and adolescent patients with SSD were implanted between 2011 and 2017. The median age at implantation was 8.9 years (range, 1.5-15.1) and the children had a median duration of deafness 2.9 years (range, 0.8-9.5). There was variability in testing measures due to patient age. Median pre-operative aided word recognition scores on the affected side were <30% regardless of the testing paradigm used. Six patients had pre-operative word testing (4 CNC, median score 25%; 2 MLNT, 8% and 17%). Four patients had pre-operative sentence testing (3 AzBio, median score 44%; 1 HINT-C, 57%). Median post-implantation follow-up interval was 12.3 months (range, 3-27.6 months). Six subjects had post-operative word recognition testing (CNC median, 70%; MLNT 50%, 92%) with a median improvement of 45.5% points. Five subjects had post-operative sentence testing (AzBio, median 82%; HINT, median 76%), with a median improvement of 40.5% points. Eight patients are full time users of their device. Tinnitus and bimodal speech reception thresholds in noise were improved.
CONCLUSION: Pediatric subjects with SSD benefit substantially from cochlear implantation. Objective speech outcome measures are improved in both quiet and noise, and bimodal speech reception thresholds in noise are greatly improved. There is a low rate of device non-use.

Zeitler DM, Dorman MF.
Cochlear Implantation for Single-Sided Deafness: A New Treatment Paradigm.
J Neurol Surg B Skull Base. 2019 Apr;80(2):178-186. doi: 10.1055/s-0038-1677482. Epub 2019 Feb 4.

ABSTRACT: Unilateral severe-to-profound sensorineural hearing loss (SNHL), also known as single sided deafness (SSD), is a problem that affects both children and adults, and can have severe and detrimental effects on multiple aspects of life including music appreciation, speech understanding in noise, speech and language acquisition, performance in the classroom and/or the workplace, and quality of life. Additionally, the loss of binaural hearing in SSD patients affects those processes that rely on two functional ears including sound localization, binaural squelch and summation, and the head shadow effect. Over the last decade, there has been increasing interest in cochlear implantation for SSD to restore binaural hearing. Early data are promising that cochlear implantation for SSD can help to restore binaural functionality, improve quality of life, and may faciliate reversal of neuroplasticity related to auditory deprivation in the pediatric population. Additionally, this new patient population has allowed researchers the opportunity to investigate the age-old question “what does a cochlear implant (CI) sound like?.”