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The Journal of Early Hearing Detection and Intervention

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What Are Others Publishing About Early Hearing Detection and Intervention?

The aim of the *Journal of Early Hearing Detection and Intervention* (JEHDI) is to promote access to evidence-based practice, standards of care, and research focused on all aspects of Early Hearing Detection and Intervention. Taking a broad systems perspective, JEHDI publishes peer-reviewed articles that describe current research, evidence-based practice, and standards of care specifically focused on newborn and early childhood hearing screening, diagnosis, support, early intervention, the medical home, information management, financing, quality improvement and other that contribute to improving EHDI systems.

Whereas *JEHDI* is the only journal that focuses specifically on improving EHDI systems, many other journals include relevant to *JEHDI*'s aim as a part their journal's broader focus. To help *JEHDI* readers stay up-to-date about recently material, we provide titles and abstracts of what has been published in the last 6 months (October 2016 through March 2017) that *JEHDI* editors think are relevant to improving EHDI programs. Titles of all articles are hyperlinked to the source.

As an indicator of what is trending in the literature, it is interesting to note that of the 73 articles published in other sources during the last 6 months:

- 11 discuss the identification or treatment of cytomegalovirus (CMV) and its relation to childhood hearing loss.
- 10 explore issues related to the genetics of hearing loss.
- 10 are about cochlear implants.
- 15 report on issues related to newborn hearing screening.
- 11 are about the diagnosis of hearing loss following screening.

Clearly, there is still much to be learned and understood about early hearing detection and intervention for infants and young children.

Abdurehim Y, Lehmann A, Zeitouni AG.
Predictive Value of GJB2 Mutation Status for Hearing Outcomes of Pediatric Cochlear Implantation.
Otolaryngol Head Neck Surg. 2017 Mar 1:194599817697054. doi:10.1177/0194599817697054. [Epub ahead of print]

Objective: To systematically review and quantify current evidence regarding the association of GJB2 mutation status with outcomes of pediatric cochlear implantation. Data Sources PubMed, Embase, and the Cochrane Library were searched for "GJB2,""pediatric hearing loss," and "cochlear implantation" and their synonyms, with no language restrictions, until December 2, 2015.

Methods: Studies were included that investigated the status of GJB2 mutation and its predictive value for outcomes of pediatric cochlear implantation. Speech recognition scores, Infant-Toddler Meaningful Auditory Integration Scale, Speech Intelligibility Rating, and Categorized Auditory Performance were pooled using weighted mean differences, and a 95% confidence interval.

Results: Eighteen studies met the inclusion criteria. The differences between GJB2-related deafness and non- GJB2-related deafness due to unidentified causes and other types of genetic deafness without additional disabilities were not statistically significant (P = .15 and P = .30, respectively); however, the difference between GJB2-related deafness and acquired hearing loss due to environmental etiologies was statistically significant and favored GJB2-related deafness (P = .03).

Conclusion: GJB2-related deafness leads to significantly better cochlear implantation outcomes when compared with acquired deafness caused by environmental etiologies. However, GJB2 mutation is not associated with a significantly better prognosis when compared with those whose deafness results from either nonsyndromic hearing loss of unknown origin or other types of genetic mutations in the absence of other neurologic deficits.

Ajalloueyan M, Saeedi M, Sadeghi M, Zamiri Abdollahi F(3).
The effects of cochlear implantation on vestibular function in 1-4 years old children.
Int J Pediatr Otorhinolaryngol. 2017 Mar;94:100-103. doi: 10.1016/j.ijporl.2017.01.019. Epub 2017 Jan 16.

Objectives: Although cochlear implants offer an effective hearing restoration option in children with severe to profound hearing loss, concern continues to exist regarding the possible effects of cochlear implantation on the vestibular system and balance.

Methods: In a prospective cohort study, 27 children with bilateral profound hearing loss (all candidates for cochlear implantation) were evaluated for their vestibular function before and after cochlear implantation. Vestibular evaluations consisted of Vestibular Evoked Myogenic Potentials, caloric testing and the Head-Impulse Test.
Results: Mean age at the time of cochlear implantation was 27.19 months. Without considering vestibular evaluation results, one of the ears was selected for surgery. Vestibular tests after surgery were not indicative of any statistically significant change in vestibular system or balance.
Conclusion: This limited data shows that cochlear implantation did not impair the vestibular system of these patients. By the results of our study we may conclude that round window implantation does not have any disturbing impact on vestibular function in children. The generalization of this result needs further research.

Amir J, Atias J, Linder N, Pardo J.
Follow-up of infants with congenital cytomegalovirus and normal fetal imaging.
Arch Dis Child Fetal Neonatal Ed. 2016 Sep;101(5):F428-32. doi:10.1136/archdischild-2015-308357. Epub 2016 Jan 18.
Objective: To evaluate the outcome of infants with congenital cytomegalovirus (CMV) infection and normal fetal imaging.
Design: Retrospective cohort study.
Setting: Tertiary paediatric medical centre.
Patients: 98 infants born to mothers with primary CMV infection in the first and second trimesters (diagnosed by positive amniotic fluid findings) and normal fetal imaging.
Methods: Initial evaluation included confirmatory urine culture, complete blood count, liver and kidney function tests, funduscopy, brain ultrasound and hearing test. Follow-up included periodic neurological and developmental evaluation, hearing tests until age 5 and Bayley-III Developmental Scale (in some patients).
Main Outcome Measures: The presence and rate of sequelae of congenital CMV.
Results: 52 (53.1%) infants received early antiviral treatment for central nervous system symptoms or signs, mainly lenticulostriatal vasculopathy on postnatal ultrasonography (88.5%). Sensorineural hearing loss was found on first examination in 16 infants (25 ears), of whom 10 also had cranial ultrasound findings; another five with late-onset hearing loss were also treated. The median follow-up time was 32 (12-83) months. Most infants with moderate and severe hearing loss were infected in the first trimester (10 vs 2, p=0.053). At the last assessment, eight children (10 ears) still had hearing loss, including two with bilateral loss who underwent a cochlear implant. The mean Bayley-III score was 102.6±10.3 (range 85-127). All 98 children attended regular educational institutions.
Conclusions: Congenital CMV infection acquired from primary maternal infection with normal fetal imaging is associated with a high rate of subtle signs and symptoms after birth. Overall, intermediate-term outcome is good with a low rate of sequelae.

Amraei K, Amirsalari S, Ajalloueyan M.
Comparison of intelligence quotients of first- and second-generation deaf children with cochlear implants.
Int J Pediatr Otorhinolaryngol. 2017 Jan;92:167-170. doi: 10.1016/j.ijporl.2016.10.005. Epub 2016 Oct 6.
Hearing impairment is a common type of sensory loss in children. Studies indicate that children with hearing impairment are deficient in social, cognitive and communication skills. This study compared the intelligence quotients of first-and second-generation deaf children with cochlear implants. This research is causal-comparative. All 15 deaf children investigated had deaf parents and were selected from Baqiyatallah Cochlear Implant Center. The 15 children with cochlear implants were paired with similar children with hearing parents using purposive sampling. The findings show that the Hotelling trace of multivariate analysis of variance ($F = 6.78$, $p < 0.01$, $\eta P(2) = 0.73$) was significant. The tests of between-subjects effects for second-generation children was significantly higher than for first-generation children for all intelligence scales except knowledge. It can be assumed that second-generation children joined their family in the use of sign language as the primary experience before a cochlear implant. The use of sign language before cochlear implants is recommended.

Amundsen VV, Wie OB, Myhrum M, Bunne M.
The impact of ethnicity on cochlear implantation in Norwegian children.
Int J Pediatr Otorhinolaryngol. 2017 Feb;93:30-36. doi: 10.1016/j.ijporl.2016.12.002. Epub 2016 Dec 3.
Objectives: To explore the impact of parental ethnicity on cochlear implantation in children in Norway with regard to incidence rates of cochlear implants (CIs), comorbidies, age at onset of profound deafness (AOD), age at first implantation, uni- or bilateral CI, and speech recognition.
Method: This retrospective cohort study included all children (N = 278) aged <18 years in Norway who received their first CI during the years 2004-2010.
Results: 86 children (30.9%) in our study sample had parents of non-Nordic ethnicity, of whom 46 were born in Nordic countries with two non-Nordic parents. Compared with the background population, children with non-Nordic parents were 1.9 times more likely to have received CI than Nordic children (i.e., born in Nordic countries with Nordic parents). When looking at AOD, uni-vs. bilateral CIs, and comorbidities, no significant differences were found between

Nordic children and children with a non-Nordic ethnicity. Among children with AOD <1 year (n = 153), those born in non-Nordic countries with two non-Nordic parents (n = 6) and adopted non-Nordic children (n = 6) received their first CI on average 14.9 and 21.1 months later than Nordic children (n = 104), respectively (p = 0.006 and 0.005). Among children with AOD <1 year, those born in Nordic countries with two non-Nordic parents (n = 31) received their CI at an older age than Nordic children, but this difference was not significant after adjusting for calendar year of implantation and excluding comorbidity as a potential cause of delayed implantation. The mean age at implantation for children with AOD <1 year dropped 2.3 months/year over the study period. The mean monosyllable speech recognition score was 84.7% for Nordic children and 76.3% for children born in Norway with two non-Nordic parents (p = 0.002). **Conclusions:** The incidence of CI was significantly higher in children with a non-Nordic vs. a Nordic ethnicity, reflecting a higher incidence of profound deafness. Children born in Norway have equal access to CIs regardless of their ethnicity, but despite being born and receiving care in Norway, prelingually deaf children with non-Nordic parents are at risk of receiving CI later than Nordic children. Moreover, prelingually deaf children who arrive in Norway at an older age may be at risk for a worse prognosis after receiving a CI due to lack of auditory stimulation in early childhood, which is critical for language development and late mplantation; this is a serious issue with regard to deafness among refugees.

Anne S, Trosman S, Haffey T, Sindwani R, Geelan-Hansen K.

Charges associated with imaging techniques in evaluation of pediatric hearing loss.

Int J Pediatr Otorhinolaryngol. 2016 Oct;89:25-7. doi: 10.1016/j.ijporl.2016.07.023. Epub 2016 Jul 25.

Objective: The best imaging study for evaluation of pediatric hearing loss is debated and it is well known magnetic resonance imaging is more costly than computed tomography. The objective of this study is to evaluate charges of computed tomography temporal bone (CTTB) versus magnetic resonance imaging brain, internal auditory canal/cerebellopontine angle (MRI IAC/CPA), with and without sedation in the pediatric population in order to assess to what extent the charges for the procedure are increased. In addition, differences in need for sedation and duration of sedation will be evaluated.

Methods: All patients, 0-18 years that underwent CTTB or MRI IAC/CPA, between January 2013 through December 2014 within the department of otolaryngology.

Results: 120 CTTBs (118 non-sedated and 2 sedated) and 51 MRI IAC/CPAs (32 non-sedated and 19 sedated) were performed. Average charge for non-sedated CTTB was $1856. CTTB scan under sedation incurred total additional charges of $2385. Average charges for non-sedated MRI IAC/CPA was $3770. Technical charges for sedated MRI IAC/CPA was $151 lower ($2858) but had additional sedation charges of $2256, a recovery room charge of $250, and additional professional fees of $1496 for total charges of $7621. 37% of MRI IAC/CPAs needed sedation to be completed in comparison to 1.6% of CTTB.

Conclusion: MRI IAC/CPAs are, on average, twice as costly as CTTBs. Almost 40% of patients need sedation to complete MRI IAC/CPA. These considerations may factor into decision making when choosing imaging modality in evaluation of pediatric hearing loss.

Ari-Even Roth D, Hildesheimer M, Roziner I, Henkin Y.

Evidence for a Right-Ear Advantage in Newborn Hearing Screening Results.

Trends Hear. 2016 Dec 6;20. pii: 2331216516681168. DOI:10.1177/2331216516681168.

The aim of the present study was to investigate the effect of ear asymmetry, order of testing, and gender on transient-evoked otoacoustic emission (TEOAE) pass rates and response levels in newborn hearing screening. The screening results of 879 newborns, of whom 387 (study group) passed screening successfully in only one ear in the first TEOAE screening, but passed screening successfully in both ears thereafter, and 492 (control group) who passed screening successfully in both ears in the first TEOAE, were retrospectively examined for pass rates and TEOAE characteristics. Results indicated a right-ear advantage, as manifested by significantly higher pass rates in the right ear (61% and 39% for right and left ears, respectively) in the study group, and in 1.75 dB greater TEOAE response amplitudes in the control group. The right-ear advantage was enhanced when the first tested ear was the right ear (76%). When the left ear was tested first, pass rates were comparable in both ears. The right-ear advantage in pass rates was similar in females versus males, but manifested in 1.5 dB higher response amplitudes in females compared with males, regardless of the tested ear and order of testing in both study and control groups. The study provides further evidence for the functional lateralization of the auditory system at the cochlear level already apparent soon after birth in both males and females. While order of testing plays a significant role in the asymmetry in pass rates, the innate right-ear advantage seems to be a more dominant contributor.

Bakhos D, Marx M, Villeneuve A, Lescanne E, Kim S, Robier A.

Electrophysiological exploration of hearing.

Eur Ann Otorhinolaryngol Head Neck Dis. 2017 Mar 16. pii: S1879-7296(17)30050-9. doi: 10.1016/j.anorl.2017.02.011. [Epub ahead of print]

Electrophysiologic hearing tests have been developed since the 1960s to determine hearing thresholds objectively. They are now implemented in newborn hearing screening. While they determine thresholds, interpretation requires subjective pure-tone and speech audiometry to determine the type of hearing loss. Each examination tests a different anatomic region, enabling the auditory system to be explored from the organ of Corti to the auditory cortex. Thus, the various objective audiometric examinations are complementary.

Bosteels S, Vandenbroeck M, Van Hove G

Saving Deaf Children? Screening for Hearing loss as a Public-interest Case.

J Bioeth Inq. 2017 Mar;14(1):109-121. doi: 10.1007/s11673-016-9752-y. Epub 2016 Oct 19.

New-born screening programs for congenital disorders and chronic disease are expanding worldwide and children "at risk" are identified by nationwide tracking systems at the earliest possible stage. These practices are never neutral and raise important social and ethical questions. An emergent concern is that a reflexive professionalism should interrogate the ever earlier interference in children's lives. The Flemish community of Belgium was among the first to generalize the screening for hearing loss in young children and is an interesting case to study the public justification of early interventions for families with deaf children. This article uses a critical lens to study the archive of the government child healthcare organization in Flanders in order to uncover underlying constructions of childhood, deafness, and preventive health. We focus on two interrelated themes. The first is the notion of exclusion of the human factor through the mediation of technology. The second is the idea of deafness as endangering a healthy development, an impairment that can nevertheless be treated if detected early enough. It is argued that, since deafness cannot be viewed as a life-threatening condition, the public interest which is implicitly defended is not the rescue of deaf children rather the exclusion of otherness.

Buxmann H, Hamprecht K, Meyer-Wittkopf M, Friese K.

Primary Human Cytomegalovirus (HCMV) Infection in Pregnancy.

Dtsch Arztebl Int. 2017 Jan 27;114(4):45-52. doi: 10.3238/arztebl.2017.0045.

Background: In 0.5-4% of pregnancies, the prospective mother sustains a primary infection with human cytomegalovirus (HCMV). An HCMV infection of the fetus in the first or second trimester can cause complex post-encephalitic impairment of the infant brain, leading to motor and mental retardation, cerebral palsy, epilepsy, retinal defects, and progressive hearing loss.

Methods: This review is based on pertinent publications from January 2000 to October 2016 that were retrieved by a selective search in PubMed employing the terms "cytomegalovirus and pregnancy" and "congenital cytomegalovirus."

Results: 85-90% of all neonates with HCMV infection are asymptomatic at birth. The main long-term sequela is hearing impairment, which develops in 8-15% of these affected children. Hygienic measures can lower the risk of primary HCMV infection in pregnancy by 50-85%. The first randomized and controlled trial (RCT) of passive immunization with an HCMV-specific hyper - immune globulin (HIG) preparation revealed a trend toward a lower risk of congenital transmission of the virus (30% versus 44% with placebo, p = 0.13). The effect of HIG was more marked in the initial non-randomized trial (15% versus 40%, p = 0.02). The RCT also showed HIG to be associated with a higher frequency of fetal growth retardation and premature birth (13% versus 2%, p = 0.06). Valaciclovir is a further, non-approved treatment option.

Conclusion: In the absence of an active vaccine against HCMV, counseling about hygienic measures may currently be the single most effective way to prevent congenital HCMV infection. Moreover, HCMV serologic testing is recommended in the guideline of the Association of the Scientific Medical Societies in Germany (Arbeitsgemeinschaft der Wissenschaftlichen Medizinischen Fachgesellschaften, AWMF). Further randomized trials of treatment with HIG and with valaciclovir are urgently needed so that the options for the prevention and treatment of congenital HCMV infection can be assessed.

Cannie MM, Devlieger R, Leyder M, Claus F, Leus A, De Catte L, Cossey V, Foulon I, Van der Valk E, Foulon W, Cos T, Bernaert A, Oyen R, Jani JC.

Congenital cytomegalovirus infection: contribution and best timing of prenatal MR imaging.

Eur Radiol. 2016 Oct;26(10):3760-9. doi: 10.1007/s00330-015-4187-0. Epub 2016 Mar 17.

Objective: To predict sensorineural hearing loss (SNHL) and neurological impairment in congenital cytomegalovirus (cCMV) infection using MR imaging and define the best timing in pregnancy for prenatal assessment.

Methods: In 121 patients with confirmed cCMV infection, brain features at MR imaging were respectively graded from

1 to 5: normal; isolated frontal/parieto-occipital hyperintensity; temporal periventricular hyperintensity; temporal/occipital cysts and/or intraventricular septa; migration disorders. Grading was correlated with postnatal SNHL and neurological impairment using regression analysis. In 51 fetuses with MR examinations at 26.9 and 33.0 weeks, the predictive value of SNHL and neurological impairment was compared using ROC curves.
Results: Postnatal follow-up showed SNHL in 18 infants and neurological impairment in 10. MR grading was predictive of SNHL and of neurological impairment ($P<0.001$). In grade 1 or 2, none had SNHL and 1/74 had neurological impairment. The areas under ROC curves for prediction of postnatal SNHL and of neurological impairment from first and second MR examination were comparable.
Conclusion: Our data suggest that in cCMV infection, prediction of SNHL and neurological impairment is feasible by fetal MR imaging with a high negative predictive value and can equally be done at 27 or 33 weeks of gestation.
Key Points: • In cCMV, isolated periventricular T2-weighted signal hyperintensity has a good postnatal prognosis. • In cCMV, SNHL and neurological impairment can be predicted at 27 or 33 weeks. • In cCMV, fetal MR has a high NPV in predicting SNHL. • In cCMV, fetal MR has a high NPV in predicting neurological impairment.

Chao X, Luo J, Fan Z, Shi H, Han Y, Wang R, Song Y, Wang G, Wang H, Xu L.
Usefulness of radiological findings for predicting cochlear implantation outcomes in children with cochlear nerve deficiency: a pilot study.
Acta Otolaryngol. 2016 Oct;136(10):1051-7. doi: 10.1080/00016489.2016.1179788. Epub 2016 May 17.
Conclusion: Children with CND received limited benefits from CIs and their results varied. The size of the vestibulocochlear nerve relative to the facial nerve could potentially be used as a predicator for CI outcomes in children with CND.
Objective: This study aimed to (1) retrospectively review the outcomes of cochlear implants (CIs) in children with cochlear nerve deficiency (CND) and (2) evaluate the clinical usefulness of radiological findings as predictors for post-implantation outcomes.
Methods: Study participants included 10 children with bilateral CND and profound sensorineural hearing loss. The preoperative magnetic resonance imaging and temporal bone computed tomography scans were evaluated. Auditory processing capability and speech perception performance were measured with Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scales. Aided hearing thresholds with CI were measured. The relationships between CI outcomes and the sizes of vestibulocochlear nerve and cochlear nerve canal (CNC) were analysed.
Results: Although post-operative CAP scores and hearing thresholds significantly improved in children with CND, their results were worse than those measured in implanted children with normal cochlear nerve. No significant correlation was found between the CI outcomes and the vestibulocochlear nerve diameters or the CNC diameters in children with CND. However, children with larger vestibulocochlear-nerve-to-facial-nerve-ratios got better results.

Chen Y, Liu Y, Wang B, Mao J, Wang T, Ye K, Ye Y, Cram DS, Li H.
Development and validation of a fetal genotyping assay with potential for noninvasive prenatal diagnosis of hereditary hearing loss.
Prenat Diagn. 2016 Dec;36(13):1233-1241. doi: 10.1002/pd.4962. Epub 2016 Dec 9.
Objective: Inherited non-syndromic hearing loss (NSHL) is a common sensory disorder that afflicts otherwise healthy individuals. The aim of the study was to evaluate the performance of circulating single molecule amplification and re-sequencing technology (cSMART) for non-invasive prenatal testing (NIPT) of NSHL.
Method: Neonatal inheritance of NSHL mutations was determined from bloodspots using SNaPshot genotyping. NIPT of cell-free DNA for fetal NSHL mutations in the GJB2, GJB3 and SLC26A4 genes was performed by a multiplex cSMART assay. The percentage of mutant alleles was used to deduce fetal DNA fractions and assign fetal genotypes.
Results: A total of 25 plasma samples selected with different fetal NSHL genotypes were coded and retrospectively analyzed by NIPT. Three normal fetuses, 18 carrier fetuses comprising seven GJB2 109G>A, four GBJ2 235delC, three GJB2 299-300delAT and four SLC26A4 IVS7-2A>G heterozygotes and four affected fetuses comprising two GJB2 109G>A homozygotes, one GBJ2 235delC homozygote and one compound GJB2 235delC/299-300delAT heterozygote were identified. All 25 fetal genotypes determined by the cSMART assay were concordant with neonatal genotypes.
Conclusion: The cSMART assay applied to cell-free DNA isolated from maternal plasma of pregnant women is highly accurate for calling correct fetal NSHL genotypes.

Cheong JP, Soo SS, Manuel AM.
Factors contributing to hearing impairment in patients with cleft lip/palate in Malaysia: A prospective study of 346 ears.
Int J Pediatr Otorhinolaryngol. 2016 Sep;88:94-7. doi: 10.1016/j.ijporl.2016.06.045. Epub 2016 Jun 29.
Objective: To determine the factors contributing towards hearing impairment in patients with cleft lip/palate.
Method: A prospective analysis was conducted on 173 patients (346 ears) with cleft lip and palate (CL/P) who presented to the combined cleft clinic at University Malaya Medical Centre (UMMC) over 12 months. The patients'

hearing status was determined using otoacoustic emission (OAE), pure tone audiometry (PTA) and auditory brainstem response (ABR). These results were analysed against several parameters, which included age, gender, race, types of cleft pathology, impact and timing of repair surgery.
Results: The patients' age ranged from 1-26 years old. They comprised 30% with unilateral cleft lip and palate (UCLP), 28% with bilateral cleft lip and palate (BCLP), 28% with isolated cleft palate (ICP) and 14% with isolated cleft lip (ICL). Majority of the patients (68.2%) had normal otoscopic findings. Out of the 346 ears, 241 ears (70%) ears had passed the hearing tests. There was no significant relationship between patients' gender and ethnicity with their hearing status. The types of cleft pathology significantly influenced the outcome of PTA and ABR screening results ($p < 0.001$). There was no significant difference between the repaired and unrepaired cleft groups and the outcome of hearing tests. However, hearing improvement occurred when palatal repair was performed at the age of <1year old (OR = 2.37, CI 1.2 = 4.6, p = 0.01).
Conclusion: Majority of the cleft patients had normal hearing (70%). Hearing threshold varied significantly between the different types of cleft pathology. Surgery conferred no significant impact on the hearing outcome unless surgery was performed at the age of <1 year old.

Chiou ST, Lung HL, Chen LS, Yen AM, Fann JC, Chiu SY, Chen HH.
Economic evaluation of long-term impacts of universal newborn hearing screening.
Int J Audiol. 2017 Jan;56(1):46-52. Epub 2016 Sep 6. DOI:10.1080/14992027.2016.1219777
Objective: Little is known about the long-term efficacious and economic impacts of universal newborn hearing screening (UNHS).
Design: An analytical Markov decision model was framed with two screening strategies: UNHS with transient evoked otoacoustic emission (TEOAE) test and automatic acoustic brainstem response (aABR) test against no screening. By estimating intervention and long-term costs on treatment and productivity losses and the utility of life years determined by the status of hearing loss, we computed base-case estimates of the incremental cost-utility ratios (ICURs). The scattered plot of ICUR and acceptability curve was used to assess the economic results of aABR versus TEOAE or both versus no screening.
Study Sample: A hypothetical cohort of 200,000 Taiwanese newborns.
Results: TEOAE and aABR dominated over no screening strategy (ICUR=$-4800.89 and $-4111.23, indicating less cost and more utility). Given $20,000 of willingness to pay (WTP), the probability of being cost-effective of aABR against TEOAE was up to 90%.
Conclusions: UNHS for hearing loss with aABR is the most economic option and supported by economically evidence-based evaluation from societal perspective.

Dar L, Namdeo D, Kumar P, Thakar A, Kant S, Rai S, Singh PK, Kabra M, Fowler KB, Boppana SB.
Congenital Cytomegalovirus Infection and Permanent Hearing Loss in Rural North Indian Children.
Pediatr Infect Dis J. 2016 Dec 28. doi: 10.1097/INF.0000000000001527. [Epub ahead of print]
Background: Congenital cytomegalovirus infection (cCMV) is a leading non-genetic cause of permanent congenital or early-onset hearing loss (PCEHL). Although cCMV rates are high despite near-universal seroimmunity, the contribution of cCMV to PCEHL in the developing world is unclear.
Methods: Neonates at a rural north Indian hospital were screened for cCMV by saliva PCR and hearing by distortion product otoacoustic emission (DPOAE) testing. CMV positive infants and those not passing newborn hearing screening (NHS) were evaluated by auditory brainstem response to confirm PCEHL. Infants with cCMV and those with PCEHL were tested for mutations within the GJB2 gene.
Results: Of the 1720 infants screened, 40 (2.3%) did not pass NHS and 20 (1.2%) were CMV positive. ABR testing confirmed unilateral or bilateral PCEHL in 11 (0.64%) children who either did not pass NHS or CMV positive. PCEHL was 20-fold higher in neonates with cCMV (2/20, 10%) than those without (9/1700, 0.5%; $p<0.01$). None of 11 infants with PCEHL had connexin 26 mutations.
Conclusion: PCEHL incidence is high in India, with cCMV contributing significantly despite near universal seroimmunity. Our findings also demonstrate the feasibility and the utility of simultaneous newborn screening for both cCMV and hearing loss in a resource-limited setting.

Diener ML, Zick CD, McVicar SB, Boettger J, Park AH.
Outcomes From a Hearing-Targeted Cytomegalovirus Screening Program.
Pediatrics. 2017 Feb;139(2). pii: e20160789. doi: 10.1542/peds.2016-0789.
Background and Objectives: Cytomegalovirus (CMV) is the most common congenital infection and nongenetic cause of congenital sensorineural hearing loss in the United States. Utah was the first state to pass legislation mandating CMV screening for newborns who fail newborn hearing screening (NBHS). The study objective was to present outcomes of hearing-targeted CMV screening and determine factors predicting CMV screening.
Methods: We used Utah Department of Health HiTrack and Vital Records databases to examine CMV screening from 509 infants who failed NBHS in the 24 months after implementation of the Utah legislation. Multivariate logistic regres-

sion analyses were conducted to identify predictors of compliance with CMV screening and diagnostic hearing evaluation.
Results: Sixty-two percent of infants who never passed hearing screening underwent CMV screening. Fourteen of 234 infants tested within 21 days were CMV positive; 6 (42.9%) had hearing loss. Seventy-seven percent of eligible infants completed a diagnostic hearing evaluation within 90 days of birth. Compliance with CMV screening was associated with sociodemographic factors, time since the law was enacted, and NBHS protocol. Infants born after the legislation showed greater odds of achieving timely diagnostic hearing evaluation than infants born before the law.
Conclusions: Incorporating CMV screening into an established NBHS program is a viable option for the identification of CMV in infants failing NBHS. The addition of CMV testing can help a NBHS program attain timely audiological diagnostics within 90 days, an important early hearing detection and intervention milestone.

Dimitriou A, Perisanidis C, Chalkiadakis V, Marangoudakis P, Tzagkaroulakis A, Nikolopoulos TP.
The universal newborn hearing screening program in a public hospital: The importance of the day of examination.
Int J Pediatr Otorhinolaryngol. 2016 Dec;91:90-93. doi:10.1016/j.ijporl.2016.10.015. Epub 2016 Oct 14.
Objectives: Newborn hearing screening programs are already implemented in many countries worldwide. Nonetheless there is still no consensus about the most proper post-birth day of examination. The purpose of this study was to assess the most appropriate day of universal hearing screening program in a public hospital.
Material and Methods: A prospective cohort study was conducted in "Attiko University National Health System Hospital" and included 2494 newborns. They were examined before discharge from the hospital, using transient evoked otoacoustic emissions (TEOAEs).
Results: From 2494 neonates included in the study, 2129 (85.4%) bilaterally passed the screening examination, while 365 (14.6%) failed the test. Higher levels of "pass" result per day of life were presented the third (90%) and fourth (94%) day of life. These days the referral scores were lower, reaching 6% the 4th post-birth day.

Driver S, Jiang D.
Paediatric cochlear implantation factors that affect outcomes.
Eur J Paediatr Neurol. 2017 Jan;21(1):104-108. doi: 10.1016/j.ejpn.2016.07.012. Epub 2016 Jul 21.
Cochlear implantation is an established surgical intervention for individuals with bilateral severe to profound sensorineural hearing loss. The aim of the intervention is to provide the individual with a sensation of sound which they can learn to interpret with meaning. Outcomes vary considerably and the factors that impact on outcomes will be discussed.

Fang X, Li X, Zhang Q, Wan J, Sun M, Chang F, Lü J, ChenG.
Universal neonatal hearing screening program in Shanghai, China: An inter-regional and international comparison.
Int J Pediatr Otorhinolaryngol. 2016 Nov;90:77-85. doi:10.1016/j.ijporl.2016.08.022. Epub 2016 Aug 29.
Objective: By comparing the Universal Neonatal Hearing Screening (UNHS) program as implemented in Shanghai and other regions in China and countries around the world, this study makes an assessment of the Shanghai model and summarizes the experiences implementing the UNHS program, so as to provide a valuable reference for other countries or regions to carry out UNHS more effectively. Since Shanghai is one of the most developed regions in China, we also examined the relationship between economic development and the UNHS starting year and coverage rate.
Methods: The study conducted a systematic review of published studies in Chinese and English on the program status of neonatal hearing screening to compare and analyze the implementation of the UNHS program in 20 cities or provinces in China and 24 regions or countries around the world. The literature search in Chinese was conducted in the three most authoritative publication databases, CNKI (China National Knowledge Infrastructure), WANFANGDATA, and CQVIP (http://www.cqvip.com/). We searched all publications in those databases with the keywords "neonatal hearing screening" (in Chinese) between 2005 and 2014. English literature was searched using the same keywords (in English). The publication database included Medline and Web of Science, and the search time period was 2000-2014.
Results: Shanghai was one of the first regions in China to implement UNHS, and its coverage rate was among the top regions by international comparison. The starting time of the UNHS program had no relationship with the Gross Domestic Product (GDP) per capita in the same year. Economic level serves as a threshold for carrying out UNHS but is not a linear contributor to the exact starting time of such a program. The screening coverage rate generally showed a rising trend with the increasing GDP per capita in China, but it had no relationship with the area's GDP per capita in selected regions and countries around the world. The system design of UNHS is the key factor influencing screening coverage. Policy makers, program administrators, and cost-sharing structures are important factors that influence the coverage rates of UNHS.
Conclusion: When to carry out a UNHS program is determined by the willingness and preference of the local

government, which is influenced by the area's social, political and cultural conditions. Mandatory hearing screening and minimal-cost to no-cost intervention are two pillars for a good coverage rate of UNHS. In terms of system design, decision-making, implementation, funding and the concrete implementation plan are all important factors affecting the implementation of the UNHS.

Farahani F, Hamidi Nahrani M, Seifrabiei MA, Emadi M.
The Effect of Mode of Delivery and Hospital Type on Newborn Hearing Screening Results Using Otoacoustic Emissions: Based on Screening Age.
Indian J Otolaryngol Head Neck Surg. 2017 Mar;69(1):1-5. doi: 10.1007/s12070-016-0967-3. Epub 2016 Feb 22.

It is well known that false positive on newborn hearing screening increases cost and maternal anxiety and worry. We aimed to evaluate the influence of mode of delivery (cesarean, vaginal) and hospital type (private, public) on false positives first screening test based on screening age. Identification and control of these factors can reduce the rate of false positives. Overall, 2784 infants were evaluated by otoacoustic emissions test. Hearing screening test was performed before hospital discharge. Finally, rate of the false-positive between both delivery group and hospital types were compared on the basis of screening age. False-positive results are obtained when a condition is not present, but the test results indicate that it is present. False positive rate in the first screening test in vaginal delivery was significantly higher than cesarean delivery and rate of significantly decreased with screening age. This reduction was observed only in cesarean delivery. Also the rate of false positives in public hospital is 2.2 fold higher than private hospital (P = 0.000) and with increase in screening age, the rate of False positive is significantly reduced in private hospitals while this decrease is not observed in public hospital. Screening test be retarded as much as possible in cesarean group and private hospital and be conducted just prior to hospital discharge also in public hospital, screening test are done in a separate room. In this way, false positive can be reduced by about six times and the cost and concerns imposed by the rate of false positives minimized.

Fowler KB, McCollister FP, Sabo DL, Shoup AG, Owen KE, Woodruff JL, Cox E, Mohamed LS, Choo DI, Boppana SB; CHIMES Study.
A Targeted Approach for Congenital Cytomegalovirus Screening Within Newborn Hearing Screening.
Pediatrics. 2017 Feb;139(2). pii: e20162128. doi: 10.1542/peds.2016-2128. Epub 2017 Jan 3. DOI:10.1542/peds.2016-2128

Background and Objective: Congenital cytomegalovirus (cCMV) infection remains a leading cause of childhood hearing loss. Currently universal CMV screening at birth does not exist in the United States. An alternative approach could be testing infants who do not pass their newborn hearing screening (NHS) for cCMV. This study was undertaken to evaluate whether a targeted approach will identify infants with CMV-related sensorineural hearing loss (SNHL).
Methods: Infants born at 7 US medical centers received NHS and were also screened for cCMV while in the newborn nursery. Infants who tested positive for CMV received further diagnostic audiologic evaluations to identify or confirm hearing loss.
Results: Between 2007 and 2012, 99 945 newborns were screened for both hearing impairment and cCMV. Overall, 7.0% of CMV-positive infants did not pass NHS compared with 0.9% of CMV-negative infants (P < .0001). Among the cCMV infants who failed NHS, diagnostic testing confirmed that 65% had SNHL. In addition, 3.6% of CMV-infected infants who passed their NHS had SNHL confirmed by further evaluation during early infancy. NHS in this cohort identified 57% of all CMV-related SNHL that occurred in the neonatal period.
Conclusions: A targeted CMV approach that tests newborns who fail their NHS identified the majority of infants with CMV-related SNHL at birth. However, 43% of the infants with CMV-related SNHL in the neonatal period and cCMV infants who are at risk for late onset SNHL were not identified by NHS.
DOI:10.1542/peds.2016-2128

Gürtler N, Gysin C, Schmid N, Pieren C, Vischer M, Schumacher S, Oppermann P, Leuba D, Veraguth D.
Bilateral congenital deafness: What investigations should be performed?
Swiss Med Wkly. 2017 Mar 21;147:w14416. doi: smw.2017.14416. eCollection 2017.

Background: The introduction of newborn hearing screening has led to earlier identification of children with congenital sensorineural hearing loss (SNHL). Aetiological clarification offers several benefits. There is currently a lack of agreement on which examinations should be recommended.
Objective: Descriptive review of the literature reporting investigations performed to establish the aetiology of congenital SNHL and comparison of the management policy in Swiss referral centres.
Methods: PubMed Search from 1985 to March 2016 with specific search terms; study selection according to inclusion/exclusion criteria; narrative analysis by use of defined criteria and question-naire.
Results: Ninety-two studies were finally included in this review. Forty studies investigated more than a single aetiology. Overall frequencies of aetiological parameters investigated were: genetic (47 studies), radiological (35), ophthalmic (35), serological (32), cardiac (25), renal (14), endocrine (12), neurological (8). Most of the studies were retrospective and various limitations such as poor population description, incomplete data or deficiencies in methodological

quality were frequently detected. The variability detected in the investigative approach chosen by Swiss referral centres reflects the heterogeneous data seen in the literature.
Conclusions: The evidence in the literature regarding an appropriate evaluation is mostly of low quality and difficult to assess owing to high heterogeneity. Nevertheless, imaging, genetic testing, neuropaediatric and ophthalmological evaluations, electrocardiograms and cytomegalovirus analysis have been identified as examinations to be included in the assessment of children with congenital SNHL. There is a need for international consensus on the various issues of such an evaluation, such as choice of investigations and diagnostic criteria.

Harris M, Terlektsi E, Kyle FE.
Literacy Outcomes for Primary School Children Who Are Deaf and Hard of Hearing: A Cohort Comparison Study.
J Speech Lang Hear Res. 2017 Mar 1;60(3):701-711. doi: 10.1044/2016_JSLHR-H-15-0403.
Purpose: In this study, we compared the language and literacy of two cohorts of children with severe-profound hearing loss, recruited 10 years apart, to determine if outcomes had improved in line with the introduction of newborn hearing screening and access to improved hearing aid technology.
Method: Forty-two children with deafness, aged 5-7 years with a mean unaided loss of 102 DB, were assessed on language, reading, and phonological skills. Their performance was compared with that of a similar group of 32 children with deafness assessed 10 years earlier and also a group of 40 children with normal hearing of similar single word reading ability.
Results: English vocabulary was significantly higher in the new cohort although it was still below chronological age. Phonological awareness and reading ability had not significantly changed over time. In both cohorts, English vocabulary predicted reading, but phonological awareness was only a significant predictor for the new cohort.
Conclusions: The current results show that vocabulary knowledge of children with severe-profound hearing loss has improved over time, but there has not been a commensurate improvement in phonological skills or reading. They suggest that children with severe-profound hearing loss will require continued support to develop robust phonological coding skills to underpin reading.

Hunter LL, Keefe DH, Feeney MP, Fitzpatrick DF.
Pressurized Wideband Acoustic Stapedial Reflex Thresholds: Normal Development and Relationships to Auditory Function in Infants.
J Assoc Res Otolaryngol. 2017 Feb;18(1):49-63. doi: 10.1007/s10162-016-0595-3. Epub 2016 Dec 7.
This study analyzed effects of pressurization on wideband acoustic stapedial-muscle reflex (ASR) tests in infants cared for in normal newborn (NN) and neonatal intensive care units (NICU). Effects of hearing-screening outcomes on ASR threshold measurements were also evaluated, and a subsequent longitudinal study established normative threshold ranges over the first year after birth. An initial experiment compared thresholds in newborns measured at ambient pressure in the ear canal and at the tympanometric peak pressure. ASR thresholds for broadband noise were higher for ears that did not pass newborn hearing screening and ASR threshold was 14 dB higher for real-ear compared to coupler conditions. Effects of pressurization were significant for ears that passed screening; thus, ASR testing in infants should be conducted at tympanometric peak pressure. ASR threshold was significantly higher for ears that referred on transient evoked otoacoustic emissions and Auditory Brainstem Response (ABR) screening tests and also for ears with conductive and sensorineural hearing loss diagnosed by ABR. Developmental ASR changes were significant over the first year for both normal and NICU infants. Wideband pressurized ASR thresholds are a clinically relevant measure of newborn hearing screening and diagnostic outcomes.

Hunter LL, Keefe DH, Feeney MP, Fitzpatrick DF, Lin L.
Longitudinal development of wideband reflectance tympanometry in normal and at-risk infants.
Hear Res. 2016 Oct;340:3-14. doi: 10.1016/j.heares.2015.12.014. Epub 2015 Dec 19.
Purpose: The goals of this study were to measure normal characteristics of ambient and tympanometric wideband acoustic reflectance, which was parameterized by absorbance and group delay, in newborns cared for in well-baby and Neonatal Intensive Care Unit (NICU) nurseries, and to characterize the normal development of reflectance over the first year after birth in a group of infants with clinically normal hearing status followed longitudinally from birth to one year of age.
Methods: Infants were recruited from a well-baby and NICU nursery, passed newborn otoacoustic emissions (OAE) and automated auditory brainstem response (ABR) tests as well as follow-up diagnostic ABR and audiometry. They were tested longitudinally for up to one year using a wideband middle ear acoustic test battery consisting of tympanometry and ambient-pressure tests. Results were analyzed for ambient reflectance across frequency and tympanometric reflectance across frequency and pressure.
Results: Wideband absorbance and group delay showed large effects of age in the first 6 months. Immature absorbance and group delay patterns were apparent in the low frequencies at birth and one month, but changed substantially to a more adult-like pattern by age 6 months for both ambient and tympanometric variables. Area and length of the ear canal estimated acoustically increased up to age 1 year. Effects of race (African American and others

compared to Caucasian) were found in combination with age effects. Mean and confidence intervals are provided for use as a normative longitudinal database for newborns and infants up to one year of age, for both well-baby and NICU infants.

Isaiah A, Lee D, Lenes-Voit F, Sweeney M, Kutz W, Isaacson B, Roland P, Lee KH.
Clinical outcomes following cochlear implantation in children with inner ear anomalies.
Int J Pediatr Otorhinolaryngol. 2017 Feb;93:1-6. doi: 10.1016/j.ijporl.2016.12.001. Epub 2016 Dec 5.

Objective: A significant proportion of children with congenital hearing loss who are candidates for cochlear implants (CIs) may have inner ear malformations (IEMs). Surgical and speech outcomes following CI in these children have not been widely reported.
Methods: The charts of children who were evaluated for a CI between 1/1/1986 and 12/31/2014 at a university-based tertiary level pediatric cochlear implant center were reviewed. Principal inclusion criteria included (i) age 1-18 years, (ii) history of bilateral severe to profound sensorineural hearing loss, and (iii)
limited benefit from binaural amplification. Exclusion criteria included (i) underlying diagnosis of neurodevelopmental disorder and (ii) lack of follow up for speech assessment if a CI was performed. The following outcome measures were reviewed: (i) imaging findings with magnetic resonance imaging or high resolution computed tomography, (ii) intraoperative complications, and (iii) speech perception categorized as the ability to perceive closed set, open set, or none.
Results: The prevalence of IEMs was 27% (102 of 381), of which 79% were bilateral. Cochlear dysplasia accounted for 30% (40 of 136) of the anomalies. Seventy-eight of the 102 patients received a CI (78%). Surgery was noted to be challenging in 24% (19 of 78), with a perilymphatic gusher being the most common intraoperative finding. Cochlear dysplasia, vestibular dysplasia and cochlear nerve hypoplasia were associated with poor speech perception (open OR closed set speech recognition scores, 0-23%), although the outcomes in children withenlarged vestibular aqueduct were similar to those of children with normal inner
ear anatomy (65%).
Conclusions: Cochlear implantation is safe in children with IEMs. However, the speech perception outcomes are notably below those of patients with normal anatomy, with the exception of when an enlarged vestibular aqueduct is present.

Januário GC, Alves CR, Lemos SM, Almeida MC, Cruz RC, Friche AA.
Health Vulnerability Index and newborn hearing screening: urban inequality.
Codas. 2016 9-10;28(5):567-574. doi: 10.1590/2317-1782/20162015182.

Purpose: To analyze the intra-urban differentials related to the outcome of the Newborn Hearing Screening (NHS) of children living in Belo Horizonte tested in a reference service using the Health Vulnerability Index (HVI).
Methods: cross-sectional study with children living in Belo Horizonte evaluated by a Newborn Hearing Screening Reference Service (NHSRS) between 2010 and 2011. The HVI of the census tract of each child was obtained by the georeferencing of their respective addresses. Multivariate analysis was conducted using the
decision tree technique, considering a statistical model for each response. A thematic map of points representing the geographic distribution of the children evaluated by the NHS program was also developed.
Results: The NHS failure rate for children living in areas with very high HVI, or without HVI data, was 1.5 times higher than that for children living in other census tracts. For children living in areas of low, medium, and high HVI, who underwent NHS after 30 days of life, the NHS failure rate was 2.1 times higher in children that presented Risk Indicator for Hearing Loss (RIHL) (17.2%) than in those who did not (8.1%). Uneven distribution was observed between areas for children that underwent the NHS and those who failed it.
Conclusion: Significant intra-urban differentials were found in Belo Horizonte, indicating correlation between health vulnerability and NHS outcomes.

Jiang ZD, Ping LL.
Reduced wave amplitudes of brainstem auditory response in high-risk babies born at 28-32 week gestation.
Brain Dev. 2016 Nov;38(10):885-892. doi: 10.1016/j.braindev.2016.05.006. Epub 2016 Jun 7.

Objective: To examine brainstem auditory electrophysiology in high-risk babies born at 28-32week gestation by analysing the amplitudes of wave components in maximum length sequence brainstem auditory evoked response (MLS BAER).
Methods: 94 preterm babies, ranging in gestation 28-32weeks, with perinatal problems (high-risk) were recruited. The amplitudes of MLS BAER wave components were studied at term age (37-42weeks postconceptional age).
Results: Compared with normal term controls, the amplitude in the high-risk preterm babies was significantly smaller at the highest click rate 910/s for wave I ($p<0.01$), at all 91-910/s for wave III (all $p<0.01$) and at 455 and 910/s ($p<0.05$ and 0.01) for wave V. Compared with age-matched low-risk preterm controls, the amplitude was significantly smaller at 455 and 910/s for wave I ($p<0.05$ and 0.05), 91-910/s for wave III ($p<0.05$-0.001), and 227-910/s ($p<0.05$ and 0.01) for wave V. No differences in the V/I and V/III amplitude ratios were found between the high-risk preterm babies and the controls.

Conclusions: The amplitudes of MLS BAER wave components, mainly more central components, were reduced in the high-risk preterm babies born at 28-32week gestation. Electrophysiological activity of the brainstem auditory neuron in such babies is depressed, mainly attributed to or related to the associated perinatal problems.

Korver AM, Smith RJ, Van Camp G, Schleiss MR, Bitner-Glindzicz MA, Lustig LR, Usami SI,
Congenital hearing loss.
Boudewyns ANNat Rev Dis Primers. 2017 Jan 12;3:16094. doi: 10.1038/nrdp.2016.94.

Congenital hearing loss (hearing loss that is present at birth) is one of the most prevalent chronic conditions in children. In the majority of developed countries, neonatal hearing screening programmes enable early detection; early intervention will prevent delays in speech and language development and has long-lasting beneficial effects on social and emotional development and quality of life. A diagnosis of hearing loss is usually followed by a search for an underlying aetiology. Congenital hearing loss might be attributed to environmental and prenatal factors, which prevail in low-income settings; congenital infections, particularly cytomegalovirus infection, are also a common risk factor for hearing loss. Genetic causes probably account for the majority of cases in developed countries; mutations can affect any component of the hearing pathway, in particular, inner ear homeostasis (endolymph production and maintenance) and mechano-electrical transduction (the conversion of a mechanical stimulus into electrochemical activity). Once the underlying cause of hearing loss is established, it might direct therapeutic decision making and guide prevention and (genetic) counselling. Management options include specific antimicrobial therapies, surgical treatment of craniofacial abnormalities and implantable or non-implantable hearing devices. An improved understanding of the pathophysiology and molecular mechanisms that underlie hearing loss and increased awareness of recent advances in genetic testing will promote the development of new treatment and screening strategies.

Krishnan LA, Van Hyfte S.
Management of unilateral hearing loss.
Int J Pediatr Otorhinolaryngol. 2016 Sep;88:63-73. doi: 10.1016/j.ijporl.2016.06.048. Epub 2016 Jun 30.

Objective: A representative sample of literature regarding unilateral hearing loss (UHL) was reviewed to provide evidence of the effects of UHL and the intervention options available for children with UHL. Considerations during the assessment and management of children with UHL are illustrated using case illustrations.
Method: Research articles published from 2013 to 2015 were searched in the PubMed database using the keywords “unilateral hearing loss”. Articles from 1950 to 2013 were included from a previous literature review on minimal hearing loss [1]. A retrospective review of charts of 14 children with UHL was also conducted.
Results: The evidence indicates that children with UHL are more likely to have structural anomalies of the inner ear; may face challenges in six different domains, and have six intervention options available. Evidence also indicates that although some children appear to exhibit no delays or difficulties, others have significant challenges, some of which continue into adulthood.
Conclusions: Children with UHL have to be treated on a case-by-case basis. Parent education regarding UHL, its effects, and all available management options is critical so they can make informed decisions. Close monitoring and good communication between professionals in different domains is crucial in order to minimize the potential negative effects of UHL.

Lanzieri TM, Chung W, Flores M, Blum P, Caviness AC, Bialek SR, Grosse SD, Miller JA, Demmler-Harrison G. Congenital Cytomegalovirus Longitudinal Study Group.
Hearing Loss in Children With Asymptomatic Congenital Cytomegalovirus Infection.
Pediatrics. 2017 Mar;139(3). pii: e20162610. doi: 10.1542/peds.2016-2610. Epub 2017 Feb 16.

Objectives: To assess the prevalence, characteristics, and risk of sensorineural hearing loss (SNHL) in children with congenital cytomegalovirus infection identified through hospital-based newborn screening who were asymptomatic at birth compared with uninfected children.
Methods: We included 92 case-patients and 51 controls assessed by using auditory brainstem response and behavioral audiometry. We used Kaplan-Meier survival analysis to estimate the prevalence of SNHL, defined as ≥25 dB hearing level at any frequency and Cox proportional hazards regression analyses to compare SNHL risk between groups.
Results: At age 18 years, SNHL prevalence was 25% (95% confidence interval [CI]: 17%-36%) among case-patients and 8% (95% CI: 3%-22%) in controls (hazard ratio [HR]: 4.0; 95% CI: 1.2-14.5; $P = .02$). Among children without SNHL by age 5 years, the risk of delayed-onset SNHL was not significantly greater for case-patients than for controls (HR: 1.6; 95% CI: 0.4-6.1; $P = .5$). Among case-patients, the risk of delayed-onset SNHL was significantly greater among those with unilateral congenital/early-onset hearing loss than those without (HR: 6.9; 95% CI: 2.5-19.1; $P < .01$). The prevalence of severe to profound bilateral SNHL among case-patients was 2% (95% CI: 1%-9%).
Conclusions: Delayed-onset and progression of SNHL among children with asymptomatic congenital cytomegalovirus infection continued to occur throughout adolescence. However, the risk of developing SNHL after age 5 years among case-patients was not different than in uninfected children. Overall, 2% of case-patients developed SNHL that was severe enough for them to be candidates for cochlear implantation.

Leal MC, Muniz LF, Ferreira TS, Santos CM, Almeida LC, Van Der Linden V, Ramos RC, Rodrigues LC, Neto SS.
Hearing Loss in Infants with Microcephaly and Evidence of Congenital Zika Virus Infection - Brazil, November 2015-May 2016.
MMWR Morb Mortal Wkly Rep. 2016 Sep 2;65(34):917-9. doi: 10.15585/mmwr.mm6534e3.

Congenital infection with Zika virus causes microcephaly and other brain abnormalities (1). Hearing loss associated with other congenital viral infections is well described; however, little is known about hearing loss in infants with congenital Zika virus infection. A retrospective assessment of a series of 70 infants aged 0-10 months with microcephaly and laboratory evidence of Zika virus infection was conducted by the Hospital Agamenon Magalhães in Brazil and partners. The infants were enrolled during November 2015-May 2016 and had screening and diagnostic hearing tests. Five (7%) infants had sensorineural hearing loss, all of whom had severe microcephaly; however, one child was tested after receiving treatment with an ototoxic antibiotic. If this child is excluded, the prevalence of sensorineural hearing loss was 5.8% (four of 69), which is similar to that seen in association with other congenital viral infections. Additional information is needed to understand the prevalence and spectrum of hearing loss in children with congenital Zika virus infection; all infants born to women with evidence of Zika virus infection during pregnancy should have their hearing tested, including infants who appear normal at birth.

Liming BJ, Carter J, Cheng A, Choo D, Curotta J, Carvalho D, Germiller JA, Hone S, Kenna MA, Loundon N, Preciado D, Schilder A, Reilly BJ, Roman S, Strychowsky J, Triglia JM, Young N, Smith RJ.
International Pediatric Otolaryngology Group (IPOG) consensus recommendations: Hearing loss in the pediatric patient.
Int J Pediatr Otorhinolaryngol. 2016 Nov;90:251-258. doi:10.1016/j.ijporl.2016.09.016. Epub 2016 Sep 15.

Objective: To provide recommendations for the workup of hearing loss in the pediatric patient.
Methods: Expert opinion by the members of the International Pediatric Otolaryngology Group.
Results: Consensus recommendations include initial screening and diagnosis as well as the workup of sensorineural, conductive and mixed hearing loss in children. The consensus statement discusses the role of genetic testing and imaging and provides algorithms to guide the workup of children with hearing loss.
Conclusion: The workup of children with hearing loss can be guided by the recommendations provided herein.

Luz I, Ribas A, Kozlowski L, Willig M, Berberian AP.
Newborn Hearing Screening in a Public Maternity Ward in Curitiba, Brazil: Determining Factors for Not Retesting.
Int Arch Otorhinolaryngol. 2016 Oct;20(4):300-304. Epub 2015 Nov 16. DOI:10.1055/s-0035-1567866

Introduction: Law 12.303/10 requires hearing screening in newborns before hospital discharge to detect possible hearing problems within the first three months after birth. If the newborn fails the test or presents signs of risk for hearing loss, it must undergo a retest and monitoring during the first year of life. In practice, this often does not happen.
Objective: To identify, in a group of mothers of children with risk factors for hearing loss, the determining reasons for non-compliance with the auditory retest.
Method: This is a cross-sectional quantitative study. For data collection, we handed a semi-structured questionnaire to 60 mothers of babies at risk for hearing loss who did not attend the hearing retest after hospital discharge. The questionnaire investigated their age, education, marital status, level of knowledge about the hearing screening, and reasons for non-compliance with the retest. We compared and analyzed data using the Chi-square test at a s ignificance level of 0.05%.
Results: Our study found that 63% of the respondents were unaware of the hearing screening and most did not receive guidance on testing during prenatal care; 30% of participants stated forgetting as the reason for not attending the retest. There was no significant relationship between age, education, and marital status regarding knowledge about the test and the non-compliance with the retest.
Conclusion: Identified as the most significant determining factors for non-compliance with the newborn hearing screening retest were the surveyed mothers' forgetting the date, and their ignorance as to the importance of retesting.

Martínez W, Torres L.
Qualitative aspects of the process of Neonatal Hearing Screening Program in Mexico evaluated from the parental perspective.
Medwave. 2016 Dec 12;16(11):e6798. doi: 10.5867/medwave.2016.11.6798.

Introduction: The Universal Newborn Hearing Screening Program in Mexico began in 2010. Its results, published in 2013 by the National Council for the Development and Inclusion of Persons with Disabilities (CONADIS), report low coverage and, currently, there is a dearth of information about its activities. This study describes the process of the program from the epistemological perspective of women whose children participated in the program, evaluating it under the sustenance of the constructivist-respondent model in search of aspects that could help explain its results.
Methods: Descriptive study with a qualitative approach based on the constructivist–respondent paradigm. We elected the 14 women who participated in the study through trial and number until theoretical saturation. After signing an informed consent form and respecting the confidentiality and anonymity, these women underwent semi-structured

interviews that were audio-recorded and transcribed as were conducted. The researchers separately analyzed and coded categories and conjointly summarized categories and subcategories. Validity and reliability were obtained through the credibility, transferability and triangulation.
Results: From the speeches, we obtained the general profile of the interviewed, evolution of their children in the program process and four categories with 15 subcategories related to the reconstruction of the process: knowledge, needs, feelings and attitudes. One was evaluated as favorable, six without agreement and eight as unfavorable. The latter refer to our own context.
Conclusions: The epistemological perspective of the interviewed women showed aspects that could help explain the low coverage of the program. Attention from public policies could improve this feature. With the establishment of the program, children with deafness are diagnosed and treated at a lower age than before the program.

Matulat P, Lepper I, Böttcher P, Parfitt R, Oswald H, Am Zehnhoff-Dinnesen A, Deuster D.
Two-Way Radio Modem Data Transfer for Newborn Hearing Screening Devices.
Telemed J E Health. 2017 Jan;23(1):49-54. doi: 10.1089/tmj.2016.0009. Epub 2016 Jun 6.
Introduction: The success of a newborn hearing screening program depends on successful tracking and follow-up to ensure that children who have had positive screening results in the first few days of life receive appropriate and timely diagnostic and intervention services. The easy availability, through a suitable infrastructure, of the data necessary for the tracking, diagnosis, and care of children concerned is a major key to enhancing the quality and efficiency of newborn hearing screening programs.
Materials and Methods: Two systems for the automated two-way transmission of newborn hearing screening and configuration data, based on mobile communication technology, for the screening devices MADSEN AccuScreen® and Natus Echo-Screen® were developed and tested in a field study. Radio modem connections were compared with conventional analogue modem transmissions from Natus Echo-Screen devices for duration, transmission rate, number of lost connections, and frequency of use.
Results: The average session duration was significantly lower with the MADSEN AccuScreen (12 s) and Natus Echo-Screen both with radio modem (15 s) than the Natus Echo-Screen with analogue modem (108 s). The transmission rate was significantly higher (898 and 1,758 vs. 181 bytes/s) for the devices with radio modems. Both radio modem devices had significantly lower rates of broken connections after initial connection (2.1 and 0.9 vs. 5.5%). An increase in the frequency of data transmission from the clinics with mobile radio devices was found.
Conclusions: The use of mobile communication technology in newborn hearing screening devices offers improvements in the average session duration, transmission rate, and reliability of the connection over analogue solutions. We observed a behavioral change in clinical staff using the new technology: the data exchange with the tracking center is more often used. The requirements for on-site support were reduced. These savings outweigh the small increase in costs for the Internet service provider.

Mehta D, Noon SE, Schwartz E, Wilkens A, Bedoukian EC, Scarano I, Crenshaw EB 3rd, Krantz ID.
Outcomes of evaluation and testing of 660 individuals with hearing loss in a pediatric genetics of hearing loss clinic.
Am J Med Genet A. 2016 Oct;170(10):2523-30. doi: 10.1002/ajmg.a.37855. Epub 2016 Aug 2.
Hearing loss is a relatively common condition in children, occurring in approximately 2 out of every 1,000 births with approximately 50% of reported diagnoses having a primary genetic etiology. Given the prevalence and genetic component of hearing loss, coupled with a trend toward early diagnosis with the institution of universal newborn hearing screening, The Genetics of Hearing Loss Clinic was established at The Children's Hospital of Philadelphia to manage the diagnosis, testing, and genetic counseling for individuals and families. This paper described a cohort of 660 individuals with a diagnosis of hearing loss evaluated between July 2008 and July 2015 in the Genetics of Hearing Loss Clinic. To elucidate the cause of hearing loss in this cohort for better management and prognostication, testing included single nucleotide polymorphism chromosomal microarray, hearing loss next generation sequencing panel, and additional clinical tests inclusive of thyroid and renal function studies, temporal bone magnetic resonance imaging, and electrocardiogram. Of those evaluated, most had bilateral sensorineural hearing loss, occurring in 489/660 (74%). Additionally, 612/660 (93%) of patients presented with a nonsyndromic form of hearing loss (no other observed clinical findings at the time of exam), of which pathogenic mutations in GJB2 were most prevalent. Of the individuals with syndromic manifestations (48/660), Usher and Waardenburg syndrome were most commonly observed. A family history of hearing loss (first degree relative) was present in 12.6% of families with available information. Through molecular analyses, clinical examination, and laboratory testing, a definitive etiologic diagnosis was established in 157/660 (23.8%) of individuals

Mena-Domínguez EA, Benito-Orejas JI, Ramírez-Cano B, Morais-Pérez D, Muñoz-Moreno MF.
High frequency tympanometry (1000Hz) in young infants and its comparison with otoacoustic emissions, otomicroscopy and 226Hz tympanometry.
Acta Otorrinolaringol Esp. 2016 Nov - Dec;67(6):306-314. doi: 10.1016/j.otorri.2016.01.001. Epub 2016 May 13.

Introduction and Objective: In the first 6 months of life, 226Hz tympanometry is considered an ineffective procedure for the diagnosis of otitis media with effusion. With the introduction of universal hearing screening, the use of high frequency 1000Hz (1kHz) tympanometry has been recommended. To optimise the diagnosis of neonatal hearing loss, we present this comparison, from the clinical point of view, of the results of 226Hz and 1kHz tympanometry in infants.
Materials and Methods: We designed a prospective study of 100 children under 9 months of age proceeding from our hearing screening program. We compare the result of tympanometry with binocular microscopy and transient evoked otoacoustic emissions.
Results: The application of transient otoacoustic emissions, otomicroscopy and 226Hz and 1kHz tympanometry has shown its usefulness in the management of otitis media with effusion of young infants, with a similar effectiveness between the 4 tests.
Conclusion: The joint use of otomicroscopy, transient otoacoustic emissions and 226Hz and 1kHz tympanometry, has allowed us to diagnose otitis media with effusion in young infants more accurately than each test separately. We recommend initial use of 1kHz tympanometry, at least in children younger than 7 months, but in the presence of hearing loss or an unclear result, 226Hz tympanometry is a good diagnostic complement.

Moodley S, Störbeck C.
Diagnostic hearing testing of infants aged 0-36 months in 3 South African provinces - Comparison of audiology records to HPCSA guidelines.
Int J Pediatr Otorhinolaryngol. 2016 Dec;91:152-158. doi: 0.1016/j.ijporl.2016.10.026. Epub 2016 Oct 26.

Introduction: Within the Early Hearing Detection and Intervention (EHDI) pathway, which includes the processes of screening, diagnosis and intervention for paediatric hearing loss, paediatric diagnostic audiology involves a battery of specific tests and procedures. International studies have highlighted a golden standard for diagnosis of paediatric hearing loss as based on the Joint Committee of Infant Hearing (2007) diagnostic guidelines, closely resembling the HPCSA diagnostic guidelines. There are limited South African studies on the processes and protocols followed in diagnostic paediatric audiology.
Objectives: This study aims to provide a comparison for how the tests used for diagnosis of paediatric hearing loss in South Africa (within both the public and private healthcare sectors) compare to the HPCSA recommended diagnostic guidelines.
Methods: A retrospective record review of paediatric clients with hearing loss (recruited through nonprobability convenience sampling) was conducted. This study is part of a longitudinal study of 711 deaf or hard of hearing children referred to the HI HOPES early intervention programme from September 2006 to December 2011. Diagnostic data from audiology reports of 117 children between 0 and 36 months were coded and analysed.
Results: Large variation was found in the tests included in the diagnostic audiology reports. For 22 children (19%) a comprehensive test battery was used. Health Professions Council of South Africa (HPCSA) recommended guidelines for diagnostic testing were not followed in any of the records analysed. Components of the HPCSA recommended test battery most frequently omitted was bone conduction testing. For both electrophysiology and behavioural testing, there was limited frequency specificity information. This exclusion of information is evidence of deficiencies in data recording and management, as well as having an effect on accuracy of classification of degree and type of hearing loss.
Conclusion: There are gaps in age-appropriate assessment protocols, which will have an effect on accurate differential diagnosis of paediatric hearing loss. Reasons for not including all testing components of the HPCSA recommended guidelines, as well as the possibility of developing guidelines more relevant to a developing world context, should be explored. There might be a need for. The impact of South African specific factors that have an effect on provision of accurate paediatric diagnostic audiology services should be determined.

Noguchi Y, Fukuda S, Fukushima K, Gyo K, Hara A, Nakashima T, Ogawa K, Okamoto M, Sato H, Usami SI, Yamasoba T, Yokoyama T, Kitamura K.
A nationwide study on enlargement of the vestibular aqueduct in Japan.
Auris Nasus Larynx. 2017 Feb;44(1):33-39. doi: 10.1016/j.anl.2016.04.012. Epub 2016 May 6.

Objective: To document the clinical features and associated pure-tone audiometry data in patients with enlargement of the vestibular aqueduct (EVA), and to identify risk factors for fluctuating hearing loss (HL) and vertigo/dizziness in EVA patients.
Methods: In this nationwide survey in Japan, a first survey sheet was mailed to 662 board-certified otolaryngology departments to identify the ones treating EVA patients. A second survey sheet, which contained solicited clinical information and the results of the hearing tests, was mailed to all facilities that reported treating EVA cases. We analyzed clinical information, including age at the time of the most recent evaluation, gender, EVA side, age at onset, initial symptoms, precipitating factors, and etiology from survey responses, and assessed 4-frequency (500, 1000, 2000,

and 4000Hz) pure-tone average (PTA) from accompanying pure-tone audiometry data. A multivariate logistic regression analysis was utilized to identify the possible risk factors for fluctuating HL and vertigo/dizziness.
Results: In total, 513 hospitals (response rate, 77.5%) responded to the first survey, and 113 reported treating patients with EVA. Seventy-nine out of the 113 hospitals (response rate 69.9%) responded to the second survey, and the data of 380 EVA patients were registered and analyzed. Of the 380 patients, 221 (58.2%) were female, suggesting female preponderance. The patient age ranged from 0 to 73 years (mean, 16.7 years; median, 13 years; interquartile range, 6-24 years). EVA was bilateral in 91.1% of the patients (346/380). The most prevalent initial symptom was HL (341/380), followed by vertigo/dizziness/imbalance (34/380). Sudden HL occurred secondary to head trauma in 5.3% of the patients and upper respiratory infection in 5.0%. Pure-tone audiometry showed profound HL (PTA >91dB) in 316 (52.0%) of the 608 ears in the 304 patients tested, and asymmetric HL, defined as >10dB, in 147 (48.4%) of the 304 patients. The mean PTA was 83.7dB (median, 91.3dB; interquartile range, 71.3-103.8dB), and the severity in PTA did not correlate with age. Multivariate logistic regression identified age ≥10 years (compared to age of 0-9 years), bilateral HL (compared to unilateral HL/normal hearing), a history of head trauma, and Pendred syndrome (compared to the other EVA-associated disorders) as significant risk factors for fluctuating HL and/or vertigo/dizziness.
Conclusion: The present nationwide survey of 380 EVA patients provided a more precise description of the clinical features, including risk factors for fluctuating HL and vertigo/dizziness.
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Núñez-Batalla F, Jáudenes-Casaubón C, Sequí-Canet JM, Vivanco-Allende A, Zubicaray-Ugarteche J, Cabanillas-Farpón R.
Aetiological diagnosis of child deafness: CODEPEH recommendations.
Acta Otorrinolaringol Esp. 2017 Jan - Feb;68(1):43-55. doi: 10.1016/j.otorri.2016.05.002. Epub 2016 Sep 16.
Important progress in the fields of molecular genetics (principally) and diagnostic imaging, together with the lack of a consensus protocol for guiding the diagnostic process after confirming deafness by neonatal screening, have led to this new work document drafted by the Spanish Commission for the Early Detection of Child Deafness (Spanish acronym: CODEPEH). This 2015 Recommendations Document, which is based on the most recent scientific evidence, provides guidance to professionals to support them in making decisions regarding aetiological diagnosis. Such diagnosis should be performed without delay and without impeding early intervention. Early identification of the causes of deafness offers many advantages: it prevents unnecessary trouble for the families, reduces health system expenses caused by performing different tests, and provides prognostic information that may guide therapeutic actions.

Núñez-Batalla F, Jáudenes-Casaubón C, Sequí-Canet JM, Vivanco-Allende A, Zubicaray-Ugarteche J.
[CODEPEH 2014 recommendations for the early detection of delayed hearing loss].
An Pediatr (Barc). 2016 Oct;85(4):215.e1-215.e6. doi: 10.1016/j.anpedi.2015.07.010. Epub 2015 Aug 12.
The latest scientific literature considers early diagnosis of deafness as key element to define the educational prognosis and inclusion of the deaf child, as advantage can be taken in the critical period of development (0-4 years). Highly significant differences exist between those deaf persons who have been stimulated early and those who have received late or inappropriate intervention. Early identification of late-onset disorders requires special attention and knowledge of all childcare professionals. Programs and additional actions beyond neonatal screening should be designed and planned in order to ensure that every child with a significant hearing loss is detected early. For this purpose, the Committee for the Early Detection of Deafness (CODEPEH) would like to highlight the need for continuous monitoring on the hearing health of children. And, for this reason, CODEPEH drafts the recommendations included in the present document.
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Obrycka A, Lorens A, Padilla García JL, Piotrowska A, Skarzynski H.
Validation of the LittlEARS Auditory Questionnaire in cochlear implanted infants and toddlers.
Int J Pediatr Otorhinolaryngol. 2017 Feb;93:107-116. doi:10.1016/j.ijporl.2016.12.024. Epub 2016 Dec 26.
Objectives: The LittlEARS Auditory Questionnaire (LEAQ) has so far been validated to assess auditory development in groups of normal-hearing children in over 20 different languages. Considering the huge variability in auditory development of CI children, especially since candidacy criteria have been relaxed, additional evidence to validate the use of LEAQ scores in this particular population is needed. The aim of this study is to provide evidence for the reliability and validity of LEAQ scores for assessing the auditory development of CI infants and toddlers based on an evaluation of LEAQ's internal structure and its relation to other variables.
Methods: The study was prospective, with sequential enrolment and within-subject repeated measures. It included 122 children with profound bilateral sensorineural hearing loss implanted at 6-22 months of age. All children were evaluated with the Polish version of LEAQ on the first day of CI activation and at each of four follow-up visits related to sound processor fitting. The study was undertaken in the light of current sychometric thinking about how assessment instruments should be validated. The main aim of the study was to obtain evidence for the validity of

interpreting LEAQ measures from CI children in terms of auditory development. First, in order to collect evidence for score reliability and validity based on LEAQ's internal structure, the psychometric properties of LEAQ scores from CI children were determined. A second step was to confirm validity by investigating the effect of concomitant variables on LEAQ scores. Correlations between LEAQ score and duration of hearing aid (HA) use, and between LEAQ score and duration of CI use, were investigated. Additionally, group differences in LEAQ scores between: 1) early and late implanted children; 2) children with long and short HA experience prior to implantation; and 3) children who showed responses over a wide frequency range from using their HAs (prior to implantation) vs those who did not.
Results: On each of the five administrations of LEAQ, the item difficulty indices increased (meaning the items became easier) and over the series they progressively increased with a range of: 0.01-0.62, 0.03-0.92, 0.09-1.00, 0.26-1.00, and 0.52-1.00. At the same time, item-total correlations were in the ranges: 0.09-0.77, 0.26-0.62, 0.00-0.65, 0.00-0.65, and 0.00-0.67. Cronbach's alpha values were above 0.80 for all administrations. A positive correlation between LEAQ score and duration of HA use, and subsequent duration of CI use (hearing experience) was found. When the children were stratified into groups according to age at cochlear implantation, duration of HA use before implantation, and audibility provided by HAs prior to implantation, the differences between the groups were reflected in both their rate of auditory development and their LEAQ score.
Conclusion: The interpretation of LEAQ scores from CI children in terms of auditory development was supported by the validity evidence of internal structure and from a logical relationship to other variables. (1) Psychometric properties -item difficulty, item-total correlations, and Cronbach's alpha values - indicate that LEAQ measures are highly consistent and reliably gauge the level of a CI child's auditory development. (2) There was a positive correlation between LEAQ scores and the duration of hearing experience with HAs and a later CI; similarly, there were significant differences between groups of children stratified according to the age at cochlear implantation, duration of HA use before implantation, and audibility provided by HAs prior to implantation, all of which demonstrate the expected relation between LEAQ score and concomitant variables.

Ogunkeyede SA, Adebola SO, Salman A, Lasisi AO.
Childhood hearing loss; a need for primary health care.
Int J Pediatr Otorhinolaryngol. 2017 Mar;94:117-120. doi: 10.1016/j.ijporl.2017.01.013. Epub 2017 Jan 11.
Introduction: Essential health care for children is the care of the ear.
Methods: A cross-sectional descriptive study of 155 children with hearing loss.
Results: A total of 155 pupils with hearing impairment and their parents were interviewed; 77(49.7%) males and 78(50.3%) females, age ranged from 6 to 15years (mean 9.11 ± 2.5 years). None of the participants had neonatal hearing screening. Parents detected the hearing loss at a mean age of 2.3 ± 1.1years. Initial care was given by community health workers and general medical practitioners, only 21 participants had otolaryngological consultation and none had audiological rehabilitation. Barriers to accessing services were financial constraints, poor awareness and non-availability of otolaryngological service for the hearing impaired in the communities.
CONCLUSION: Hearing impaired children in Nigeria have poor access to ear care. There is a need to create awareness of otological services and incorporate ear-care into the primary health care.
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Palabiyik FB, Hacikurt K, Yazici Z.
Facial nerve anomalies in paediatric cochlear implant candidates: radiological evaluation.
J Laryngol Otol. 2017 Jan;131(1):26-31. Epub 2016 Dec 5.
Background: Pre-operative radiological identification of facial nerve anomalies can help prevent intra-operative facial nerve injury during cochlear implantation. This study aimed to evaluate the incidence and configuration of facial nerve anomalies and their concurrence with inner-ear anomalies in cochlear
implant candidates.
Methods: Inner-ear and concomitant facial nerve anomalies were evaluated by magnetic resonance imaging and temporal high-resolution computed tomography in 48 children with congenital sensorineural hearing loss who were cochlear implant candidates.
Results: Inner-ear anomalies were present in 11 out of 48 patients (23 per cent) and concomitant facial nerve anomalies were present on 7 sides in 4 patients (7 per cent of the total). Facial nerve anomalies were accompanied by cochlear or vestibular malformation.
Conclusion: Potential facial nerve abnormalities should always be considered in patients with inner-ear anomalies. Pre-operative facial nerve imaging can increase the surgeon's confidence to plan and perform cochlear implantation. Magnetic resonance imaging should be used to detect inner-ear anomalies; if these
are identified, temporal high-resolution computed tomography should be used to evaluate the facial nerve.

Pater JA, Benteau T, Griffin A, Penney C, Stanton SG, Predham S, Kielley B, Squires J, Zhou J, Li Q, Abdelfatah N, O'Rielly DD, Young TL.
A common variant in CLDN14 causes precipitous, prelingual sensorineural hearing loss in multiple families due to founder effect.
Hum Genet. 2017 Jan;136(1):107-118. doi: 10.1007/s00439-016-1746-7. Epub 2016 Nov 12.

Genetic isolates provide unprecedented opportunities to identify pathogenic mutations and explore the full natural history of clinically heterogeneous phenotypes such as hearing loss. We noticed a unique audioprofile, characterized by prelingual and rapid deterioration of hearing thresholds at frequencies >0.5 kHz in several adults from unrelated families from the island population of Newfoundland. Targeted serial Sanger sequencing of probands for deafness alleles (n = 23) that we previously identified in this founder population was negative. Whole exome sequencing in four members of the largest family (R2010) identified a CLDN14 (DFNB29) variant [c.488C>T; p. (Ala163Val)], likely pathogenic, sensorineural hearing loss, autosomal recessive. Although not associated with deafness or disease, CLDN14 p.(Ala163Val) has been previously reported as a variant of uncertain significance (VUS). Targeted sequencing of 169 deafness probands identified one homozygote and one heterozygous carrier. Genealogical studies, cascade sequencing and haplotype analysis across four unrelated families showed all subjects with the unique audioprofile (n = 12) were also homozygous for p.(Ala163Val) and shared a 1.4 Mb DFNB29-associated haplotype on chromosome 21. Most significantly, sequencing 175 population controls revealed 1% of the population are heterozygous for CLDN14 p.(Ala163Val), consistent with a major founder effect in Newfoundland. The youngest CLDN14 [c.488C>T; p.(Ala163Val)] homozygote passed newborn screening and had normal hearing thresholds up to 3 years of age, which then deteriorated to a precipitous loss >1 kHz during the first decade. Our study suggests that genetic testing may be necessary to identify at-risk children in time to prevent speech, language and developmental delay.

Peng Q, Huang S, Liang Y, Ma K, Li S, Yang L, Li W, Ma Q, Liu Q, Zhong B, Lu X.
Concurrent Genetic and Standard Screening for Hearing Impairment in 9317 Southern Chinese Newborns.
Genet Test Mol Biomarkers. 2016 Oct;20(10):603-608. Epub 2016 Aug 19.

Objective: The goal of this study was to investigate the use of concurrent genetic screening together with standard newborn hearing screening (NHS) in an effort to provide a scientific basis for the beneficial use of concurrent genetic hearing screening in newborns. Our aim was to improve the neonatal detection rate of hearing impairment and the potential for hearing loss, allowing for increased early intervention and potentially allowing for prevention of later onset hearing loss. This information could also be used to increase the effectiveness of genetic counseling regarding hearing impairment.
Methods: A total of 9317 neonates from Children's Hospital of Dongguan and Dongguan People's Hospital were included in this study between January 2015 and October 2015. Twenty hotspot hearing-associated mutations of four common deafness- susceptibility genes (GJB2, GJB3, SLC26A4, and MTRNR1) were analyzed by matrix-assisted laser desorption-ionization time-of-flight mass spectrometry (MALDI-TOF-MS). The results of genetic screening and NHS were concurrently analyzed.
Results: A total of 129 infants (1.38%) exhibited hearing loss as determined by otoacoustic emission (OAE) testing. The genetic screening revealed that 348 (3.74%) individuals had at least one mutant allele. In total, 34 (0.36%) of the neonates carried a causal complement of mutations. The overwhelming majority of the genetically referred newborns passed the OAE hearing screening, but could be at risk for later hearing loss.
Conclusion: This study furthers the understanding of the etiology of hearing loss and proves that it is beneficial to use genetic screening along with OAE screening of neonates to improve detection rates of at-risk infants. Our results show that this concurrent testing allows for better early identification of infants at risk for hearing loss, which may occur before speech and language development. Prevention of hearing loss can be achieved by avoiding the use of antibiotics containing amino glycosides in infants whose mutations make them extremely sensitive to these antibiotics. This information is also useful in genetic counseling, providing region-specific mutation information.

Rawlinson WD, Boppana SB, Fowler KB, Kimberlin DW, Lazzarotto T, Alain S, Daly K, Doutré S, Gibson L, Giles ML, Greenlee J, Hamilton ST, Harrison GJ, Hui L, Jones CA, Palasanthiran P, Schleiss MR, Shand AW, van Zuylen WJ.
Congenital cytomegalovirus infection in pregnancy and the neonate: consensus recommendations for prevention, diagnosis, and therapy.
Lancet Infect Dis. 2017 Mar 10. pii: S1473-3099(17)30143-3. doi: 10.1016/S1473-3099(17)30143-3. [Epub ahead of print]

Congenital cytomegalovirus is the most frequent, yet under-recognised, infectious cause of newborn malformation in developed countries. Despite its clinical and public health importance, questions remain regarding the best diagnostic methods for identifying maternal and neonatal infection, and regarding optimal prevention and therapeutic strategies for infected mothers and neonates. The absence of guidelines impairs global efforts to decrease the effect of congenital cytomegalovirus. Data in the literature suggest that congenital cytomegalovirus infection remains a research

priority, but data are yet to be translated into clinical practice. An informal International Congenital Cytomegalovirus Recommendations Group was convened in 2015 to address these questions and to provide recommendations for prevention, diagnosis, and treatment. On the basis of consensus discussions and a review of the literature, we do not support universal screening of mothers and the routine use of cytomegalovirus immunoglobulin for prophylaxis or treatment of infected mothers. However, treatment guidelines for infected neonates were recommended. Consideration must be given to universal neonatal screening for cytomegalovirus to facilitate early detection and intervention for sensorineural hearing loss and developmental delay, where appropriate. The group agreed that education and prevention strategies for mothers were beneficial, and that recommendations will need continual updating as further data become available.

Ribeiro GE, Silva DP, Montovani JC.
Transient evoked otoacoustic emissions and auditory brainstem response in infants with perinatal asphyxia.
Int J Pediatr Otorhinolaryngol. 2016 Oct;89:136-9. doi: 10.1016/j.ijporl.2016.08.009. Epub 2016 Aug 15.

Objective: The objective of this study was to verify the effects of perinatal asphyxia on different parts of the auditory system.
Methods: This was a non-concurrent cohort study conducted on a fixed population in a tertiary public hospital. Participants included 181 infants born at term who underwent the transient evoked otoacoustic emission test as a part of a neonatal hearing screening program, with a "pass" result in both ears, and by auditory brainstem response testing. The infants were divided into 3 groups: G1, 20 infants who had perinatal asphyxia; G2, 111 infants with an Apgar score lower than 4 in the first minute and/or lower than 6 in the fifth minute (called "low Apgar" at birth); and G3, 50 infants with first- and fifth-minute Apgar scores ≥7.
Results: The signal-to-noise ratio of transient evoked otoacoustic emissions were greater in G3 compared with G1 and G2 at 4 kHz frequency for males. An increased latency of waves I and III in the auditory brainstem response of male infants in G1 was observed.
Conclusion: This study demonstrated that alterations occurred in both the cochlear and the neural components in male infants who had perinatal asphyxia.

Ronchi A, Shimamura M, Malhotra PS, Sánchez PJ.
Encouraging postnatal cytomegalovirus (CMV) screening: the time is NOW for universal screening!
Expert Rev Anti Infect Ther. 2017 May;15(5):417-419. doi: 10.1080/14787210.2017.1303377. Epub 2017 Mar 13.

The time for universal CMV screening is NOW! Although both targeted and universal CMV screening has been shown to be cost-effective, universal screening provides larger net savings and the greatest opportunity for directed care. The prevalence of congenital CMV infection, its associated sequelae, the availability of a simple saliva screening tool, available antiviral treatment, and directed therapies for hearing impairment mandate that we act now to make universal screening a reality!

Ross SA, Ahmed A, Palmer AL, Michaels MG, Sánchez PJ, Stewart A, Bernstein DI, Feja K, Fowler KB, Boppana SB; CMV and Hearing Multicenter Screening (CHIMES) Study Group.
Newborn Dried Blood Spot Polymerase Chain Reaction to Identify Infants with Congenital Cytomegalovirus-Associated Sensorineural Hearing Loss.
J Pediatr. 2017 May;184:57-61.e1. doi: 10.1016/j.jpeds.2017.01.047. Epub 2017 Feb 22.

Objective: To determine the utility of dried blood spot (DBS) polymerase chain reaction (PCR) in identifying infants with cytomegalovirus (CMV) infection-associated sensorineural hearing loss (SNHL).
Study Design: Newborns at 7 US hospitals between March 2007 and March 2012 were screened for CMV by saliva rapid culture and/or PCR. Infected infants were monitored for SNHL during the first 4 years of life to determine sensitivity, specificity, and positive and negative likelihood ratios of DBS PCR for identifying CMV-associated SNHL.
Results: DBS at birth was positive in 11 of 26 children (42%) with SNHL at age 4 years and in 72 of 270 children (27%) with normal hearing (P = .11). The sensitivity (42.3%; 95% CI, 23.4%-63.1%) and specificity (73.3%; 95% CI, 67.6%-78.5%) was low for DBS PCR in identifying children with SNHL at age 4 years. The positive and negative likelihood ratios of DBS PCR positivity to detect CMV-associated SNHL at age 4 years were 1.6 (95% CI, 0.97-2.6) and 0.8 (95% CI, 0.6-1.1), respectively. There was no difference in DBS viral loads between children with SNHL and those without SNHL.
Conclusions: DBS PCR for CMV has low sensitivity and specificity for identifying infants with CMV-associated hearing loss. These findings, together with previous reports, demonstrate that DBS PCR does not identify either the majority of CMV-infected newborns or those with CMV-associated SNHL early in life.

Rouillon I, Parodi M, Denoyelle F, Loundon N.
How to perform ABR in young children.
Eur Ann Otorhinolaryngol Head Neck Dis. 2016 Dec;133(6):431-435. doi:10.1016/j.anorl.2016.05.004. Epub 2016 Jul 21.
The diagnosis of hearing loss, especially in the context of newborn hearing screening, is mostly based on auditory brainstem response (ABR). According to the official CCAM nomenclature, ABR consists of recording early auditory evoked potentials to detect thresholds, study conduction times and measure amplitudes (corresponding to codes CDQP006 when performed without general anesthesia, and CDQP014 when performed with general anesthesia). ABR must be rigorously performed and interpreted, always in combination with a complete ENT examination and behavioral audiometry as soon as possible. In order to obtain good quality recordings, ABR must be performed with the infant totally immobile, during a nap. Several protocols can be used according to the child's age in order to obtain good quality sleep. ABR contribute to a precise hearing diagnosis, allowing early management by the first months of life.

Shang Y, Hao W, Gao Z, Xu C, Ru Y, Ni D.
An effective compromise between cost and referral rate: A sequential hearing screening protocol using TEOAEs and AABRs for healthy newborns.
Int J Pediatr Otorhinolaryngol. 2016 Dec;91:141-145. doi:10.1016/j.ijporl.2016.10.025. Epub 2016 Oct 26.
Objective: This study evaluated the efficacy of a sequential hearing screening protocol using transient evoked otoacoustic emission (TEOAE) and automatedauditory brainstem response (AABR) tests in healthy newborns.
Design: A TEOAE screening was performed during the first 48-72 h of life. If the infants failed, an AABR test was performed at the same time, and they were referred for a TEOAE rescreening at six weeks old. The results of s creening Protocol 1 (only TEOAE) were compared with those of screening Protocol 2 (sequential TEOAE + AABR screenings for the first screening and TEOAE for the rescreening).
Study Sample: A total of 1062 healthy newborns were enrolled in this research.
Results: For Protocol 1, the first screening and rescreening referral rates were 11.1% and 2.2%, respectively. In contrast, for Protocol 2, the referral rates were significant lower at 3.8% and 0.9%, respectively. Using the two protocols, six infants were diagnosed with hearing loss (0.57%).
Conclusions: Adding simultaneous AABR tests for infants who fail TEOAE testing at the first screening stage can significantly reduce referral rates without increasing misdiagnosis rates. Although this sequential screening process involves slightly more time and has a higher cost than TEOAE alone, its greater accuracy compensates for this difference.

Shetty HN, Koonoor V.
Sensory deprivation due to otitis media episodes in early childhood and its effect at later age: A psychoacoustic and speech perception measure.
Int J Pediatr Otorhinolaryngol. 2016 Nov;90:181-187. doi: 10.1016/j.ijporl.2016.09.022. Epub 2016 Sep 19.
Background: Past research has reported that children with repeated occurrences of otitis media at an early age have a negative impact on speech perception at a later age. The present study necessitates documenting the temporal and spectral processing on speech perception in noise from normal and atypical groups.
Objectives: The present study evaluated the relation between speech perception in noise and temporal; and spectral processing abilities in children with normal and atypical groups.
Methods: The study included two experiments. In the first experiment, temporal resolution and frequency discrimination of listeners with normal group and three subgroups of atypical groups (had a history of OM) a) less than four episodes b) four to nine episodes and c) More than nine episodes during their chronological age of 6 months to 2 years) were evaluated using measures of temporal modulation transfer function and frequency discrimination test. In the second experiment, SNR 50 was evaluated on each group of study participants. All participants had normal hearing and middle ear status during the course of testing.
Results: Demonstrated that children with atypical group had significantly poorer modulation detection threshold, peak sensitivity and bandwidth; and frequency discrimination to each F0 than normal hearing listeners. Furthermore, there was a significant correlation seen between measures of temporal resolution; frequency discrimination and speech perception in noise. It infers atypical groups have significant impairment in extracting envelope as well as fine structure cues from the signal.
Conclusion: The results supported the idea that episodes of OM before 2 years of agecan produce periods of sensory deprivation that alters the temporal and spectral skills which in turn has negative consequences on speech perception in noise.

Simonazzi G, Cervi F, Zavatta A, Pellizzoni L, Guerra B, Mastroroberto M, Morselli-Labate AM, Gabrielli L, Rizzo N, Lazzarotto T.

Congenital Cytomegalovirus Infection: Prognostic Value of Maternal DNAemia at Amniocentesis.

Clin Infect Dis. 2017 Jan 15;64(2):207-210. doi: 10.1093/cid/ciw700. Epub 2016 Oct 19.

Background: Human Cytomegalovirus (HCMV) is the most common cause of childhood hearing loss and can lead to neurodevelopmental delay. To date, few studies have examined the correlation between maternal viremia and congenital HCMV infection. The aim of our study was to ascertain if HCMV DNA in the peripheral blood of pregnant women with primary HCMV infection at the time of amniocentesis may have a prognostic value in terms of congenital infection and neonatal symptomatic disease.

Methods: We performed a prospective observational study of pregnant women referred to our maternal-fetal medicine division with suspected HCMV infection. Primary infection was diagnosed based on seroconversion for HCMV and/or HCMV immunoglobulin M-positive and low or moderate HCMV immunoglobulin G avidity. At the time of amniocentesis, maternal blood samples were collected and analyzed by means of real-time polymerase chain reaction to determine the presence of viral DNAemia. Fetuses and newborns were evaluated for the presence of congenital infection and symptomatic disease.

Results: A total of 239 pregnant women were enrolled; 32 blood samples (13.4%) were positive, and 207 (86.6%) were negative for HCMV DNA. The overall rate of transmission was 23.4%. Fifteen infected patients (26.8%) were symptomatic. Vertical transmission occurred in 14 women (43.8%) with positive and 42 (20.3%) with negative results for HCMV DNAemia (P = .006; odds ratio, 3.06; 95% confidence interval, 1.41-6.64). Symptomatic infection occurred in 6 (42.9%) infected fetuses or newborns from women with and in 9 (21.4%) from women without viral DNAemia (P = .16).

Conclusion: Maternal viremia at amniocentesis is associated with a 3-fold greater chance of congenital infection, but it is not correlated with symptomatic disease.

Sivam SK, Syms CA 3rd, King SM, Perry BP.

Consideration for routine outpatient pediatric cochlear implantation: A retrospective chart review of immediate post-operative complications.

Int J Pediatr Otorhinolaryngol. 2017 Mar;94:95-99. doi: 10.1016/j.ijporl.2016.12.018. Epub 2016 Dec 26.

Introduction: Cochlear implantation is well accepted as the treatment of choice for prelingual deafness in children [1]. However, the safety of routinely performing this procedure on an outpatient basis is debated. We aim to assess immediate postoperative complications that would affect a surgeon's decision to perform pediatric cochlear implantation on an outpatient basis.

Methods: A retrospective chart review was conducted which included all children 17 years old or younger who underwent cochlear implantation from 2004 to 2014 in a private neurotology practice. The immediate postoperative complication rates and types of complications were then examined.

Results: A total of 579 cochlear implants were placed in children ages 1-17 years old from 2004 to 2014. The most common complications were nausea/vomiting and dizziness/imbalance. The odds ratio of developing complications in the group ages 1-3 years old versus all other age patients was found to be statistically insignificant (OR 0.90, 95% CI 0.61 to 1.32, p = 0.58). The odds ratio of developing a complication after bilateral implantation compared to unilateral implantation was statistically significant (OR 1.96, 95% CI 1.18 to 3.28, p = 0.01). There was no difference in complication rates when comparing lateral wall and perimodiolar insertions. A total of 6 of 579 (1%) cochlear implants resulted in a complication requiring unplanned medical attention.

Conclusions: Overall, this series offers a decade of experience in pediatric cochlear implantation that shows a low incidence of the need for unplanned medical attention in the immediate postoperative period. The most common complication seen is Post-operative nausea and vomiting (PONV) that appears to be amenable to outpatient management even in the youngest populations. This supports providers routinely performing pediatric cochlear implantation on an outpatient basis.

Vo QT, Pham D, Choi KJ, Nguyen UT, Le L, Shanewise T, Tran L, Nguyen N, Lee WT.

Solar-powered hearing aids for children with impaired hearing in Vietnam: a pilot study.

Paediatr Int Child Health. 2017 Jan 25:1-6. doi: 10.1080/20469047.2016.1276119.

[Epub ahead of print]

Background: Hearing loss is a barrier to speech and social and cognitive development. This can be especially pronounced in children living in low- and middle-income countries with limited resources.

Aim: To determine the feasibility, durability and social impact of ComCare GLW solar-powered hearing aids provided for Vietnamese children with hearing impairment.

Methods: A retrospective review of data from an international, multi-discipline humanitarian visit was performed. Hearing aids were given to 28 children enrolled at the Khoai Chau Functional Rehabilitation School, Hung Yen

Province, Vietnam. Device inspection and observational assessments were performed by teachers using a modified Parents' Evaluation of Aural/Oral Performance of Children and an Infant Hearing Program Amplification Benefit Questionnaire. Qualitative interviews were undertaken to assess the study aims.
Results: Hearing aids were well tolerated for use during regular school hours. All units remained functional during the study period (12 months). Teachers noted increased student awareness and responsiveness to surrounding sounds, but the degree of response to amplification varied between children. There was no significant improvement in speech development as all subjects had prelingual deafness. Teachers felt confident in troubleshooting any potential device malfunction.
Conclusion: A solar-powered hearing aid may be a viable option for children in low- and middle-income countries. This study demonstrates that device distribution, maintenance and function can be established in countries with limited resources, while providing feasibility data to support future studies investigating how similar devices may improve the quality of life of those with hearing loss.

Vohr BR.
Language and hearing outcomes of preterm infants.
Semin Perinatol. 2016 Dec;40(8):510-519. doi: 10.1053/j.semperi.2016.09.003. Epub 2016 Nov 3.
Multiple factors including degree of prematurity, neonatal morbidities, illness severity, hearing status, gender, language environment in the neonatal intensive care unit and in the home, maternal education level, social and environmental status of the family, and access to early intervention all contribute to the language outcomes of extremely preterm infants with and without hearing loss. Early screening, early diagnosis, and early intervention services by 6 months of age are necessary to optimize the language outcomes of preterm infants with permanent hearing loss. There is increasing evidence of the potential for improved language skills with increasing age of extreme preterm infants and infants with hearing loss.

Voss SE, Herrmann BS, Horton NJ, Amadei EA, Kujawa SG.
Reflectance Measures from Infant Ears With Normal Hearing and Transient Conductive Hearing Loss.
Ear Hear. 2016 Sep-Oct;37(5):560-71. doi: 10.1097/AUD.0000000000000293.
Objective: The objective is to develop methods to utilize newborn reflectance measures for the identification of middle-ear transient conditions (e.g., middle-ear fluid) during the newborn period and ultimately during the first few months of life. Transient middle-ear conditions are a suspected source of failure to pass a newborn hearing screening. The ability to identify a conductive loss during the screening procedure could enable the referred ear to be either (1) cleared of a middle-ear condition and recommended for more extensive hearing assessment as soon as possible, or (2) suspected of a transient middle-ear condition, and if desired, be rescreened before more extensive hearing assessment.
Design: Reflectance measurements are reported from full-term, healthy, newborn babies in which one ear referred and one ear passed an initial auditory brainstem response newborn hearing screening and a subsequent distortion product otoacoustic emission screening on the same day. These same subjects returned for a detailed follow-up evaluation at age 1 month (range 14 to 35 days). In total, measurements were made on 30 subjects who had a unilateral refer near birth (during their first 2 days of life) and bilateral normal hearing at follow-up (about 1 month old). Three specific comparisons were made: (1) Association of ear's state with power reflectance near birth (referred versus passed ear), (2) Changes in power reflectance of normal ears between newborn and 1 month old (maturation effects), and (3) Association of ear's newborn state (referred versus passed) with ear's power reflectance at 1 month. In addition to these measurements, a set of preliminary data selection criteria were developed to ensure that analyzed data were not corrupted by acoustic leaks and other measurement problems.
Results: Within 2 days of birth, the power reflectance measured in newborn ears with transient middle-ear conditions (referred newborn hearing screening and passed hearing assessment at age 1 month) was significantly greater than power reflectance on newborn ears that passed the newborn hearing screening across all frequencies (500 to 6000 Hz). Changes in power reflectance in normal ears from newborn to 1 month appear in approximately the 2000 to 5000 Hz range but are not present at other frequencies. The power reflectance at age 1 month does not depend significantly on the ear's state near birth (refer or pass hearing screening) for frequencies above 700 Hz; there might be small differences at lower frequencies.
Conclusions: Power reflectance measurements are significantly different for ears that pass newborn hearing screening and ears that refer with middle-ear transient conditions. At age 1 month, about 90% of ears that referred at birth passed an auditory brainstem response hearing evaluation; within these ears the power reflectance at 1 month did not differ between the ear that initially referred at birth and the ear that passed the hearing screening at birth for frequencies above 700 Hz. This study also proposes a preliminary set of criteria for determining when reflectance measures on young babies are corrupted by acoustic leaks, probes against the ear canal, or other measurement problems. Specifically proposed are "data selection criteria" that depend on the power reflectance, impedance magnitude, and impedance angle. Additional data collected in the future are needed to improve and test these proposed criteria.

Wachtlin B, Brachmaier J, Amann E, Hoffmann V, Keilmann A.
Development and evaluation of the LittlEARS(®) Early Speech Production Questionnaire - LEESPQ.
Int J Pediatr Otorhinolaryngol. 2017 Mar;94:23-29. doi:10.1016/j.ijporl.2017.01.007. Epub 2017 Jan 9.

Objective: Universal Newborn Hearing Screening programs, now instituted throughout the German-speaking countries, allow hearing loss to be detected and treated much earlier than ever before. With this earlier detection, arises the need for tools fit for assessing the very early speech and language production development of today's younger (0-18 month old) children. We have created the LittlEARS(®) Early Speech Production Questionnaire, with the aim of meeting this need.
Methods: 600 questionnaires of the pilot version of the LittlEARS(®) Early Speech Production Questionnaire were distributed to parents via pediatricians' practices, day care centers, and personal contact. The completed questionnaires were statistically analyzed to determine their reliability, predictive accuracy, internal consistency, and to what extent gender or unilingualism influenced a child's score. Further, a norm curve was generated to plot the children's increased expected speech production ability with age.
Results: Analysis of the data from the 352/600 returned questionnaires revealed that scores on LittlEARS(®) Early Speech Production Questionnaire correlate positively with a child's age, with older children scoring higher than do younger children. Further, the questionnaire has a high measuring reliability, high predictability, high unidemensionality of scale, and is not significantly gender or uni-/multilingually biased. A norm curve for expected development with age was created.
Conclusions: The LittlEARS(®) Early Speech Production Questionnaire (LEESPQ) is a valid tool for assessing the most important milestones in very early development of speech and language production of German language children with normal hearing aged 0-18 months old. The questionnaire is a potentially useful tool for long-term infant screening and follow-up testing and for children with normal hearing and those who would benefit from or use hearing devices.

Walker RE, Bartley J, Flint D, Thompson JM, Mitchell EA.
Determinants of chronic otitis media with effusion in preschool children: a case-control study.
BMC Pediatr. 2017 Jan 6;17(1):4. doi: 10.1186/s12887-016-0767-7.

Background: Chronic otitis media with effusion (COME) is a prevalent upper airway infection resulting in hearing loss. The aim of this research was to determine risk factors for COME in preschool children.
Methods: A case-control design was conducted in Auckland, New Zealand from May 2011 until November 2013. The cases were children aged 3 and 4 years referred for tympanostomy tube placement due to a diagnosis of COME (n = 178). The controls were a random sample of healthy children aged 3 and 4 years from primary care practices (n = 209). The children's guardians completed an interviewer-administered questionnaire that covered topics including socio-demographic information, pregnancy and birth, infant feeding practices, home environment, and respiratory health. In addition, skin prick tests for atopy were performed. Odds ratios (OR) estimating the risk of COME independently associated with the exposures were calculated using a logistic regression model.
Results: Children with COME frequently had nasal obstruction (OR: 4.38 [95% CI: 2.37-8.28]), always snored (OR: 3.64 [95% CI: 1.51-9.15]) or often snored (OR: 2.45 [95% CI: 1.04-5.96]), spent more hours per week in daycare (OR per hour/week: 1.03 [95% CI: 1.00-1.05]), had frequent colds (OR: 2.67 [95% CI: 1.59-4.53]), had siblings who had undergone tympanostomy tube placement (OR: 2.68 [95% CI: 1.22-6.02]), underwent long labour (OR: 2.59 [95% CI: 1.03-6.79]), and had early introduction of cow's milk (OR: 1.76 [95% CI: 1.05-2.97]). Asian ethnicity (OR: 0.20 [95% CI: 0.07-0.53]) and having older siblings (OR: 0.54 [95% CI: 0.31-0.93]) were inversely associated with COME.
Conclusion: COME in preschool children was associated with pathogen exposure, respiratory infection, and nasal obstruction. Strategies to prevent pathogen transmission warrant investigation. The novel findings of long labour and early cow's milk introduction require replication in future studies.

Wang S, Wang T, Zhang W, Liu X, Wang X, Wang H, He X, Zhang S, Xu S, Yu Y, Jia X, Wang M, Xu A, Ma W, Amin MM, Bialek SR, Dollard SC, Wang C.
Cohort study on maternal cytomegalovirus seroprevalence and prevalence and clinical manifestations of congenital infection in China.
Medicine (Baltimore). 2017 Feb;96(5):e6007. doi: 10.1097/MD.0000000000006007.

Congenital cytomegalovirus (CMV) infection is the leading viral cause of birth defects and developmental disabilities in developed countries. However, CMV seroprevalence and burden of congenital CMV infection are not well defined in China.Cohort of newborns from 5 birthing hospitals in 2 counties of Shandong Province, China, were enrolled from March 2011 to August 2013. Dried blood spots (DBS) and saliva were collected within 4 days after birth for IgG testing for maternal seroprevalence and real-time PCR testing for congenital CMV infection, respectively.Among 5020 newborns tested for CMV IgG, 4827 were seropositive, resulting in CMV maternal seroprevalence of 96.2% (95% confidence interval [CI]:95.6%-96.7%). Of the 10,933 newborns screened for congenital CMV infection, 75 had CMV detected, resulting in an overall prevalence of 0.7% (95% CI: 0.5%-0.9%), with prevalences of 0.4% (14/3995), 0.6% (66/10,857), and 0.7% (52/7761) for DBS, wet saliva, and dried saliva specimens screened, respectively. Prevalence

of congenital CMV infection decreased with increasing maternal age (0.9%, 0.6%, and 0.3% among newborns delivered from mothers aged 16-25, 26-35, and >35 years, respectively; P=0.03), and was higher among preterm infants than full term infants (1.3% vs 0.6%, P=0.04), infants with intrauterine growth restriction (IUGR) than those without (1.8% vs 0.7%, P=0.03), and twins or triplets than singleton pregnancies (2.8% vs 0.7%, P=0.04). None of the 75 newborns exhibited symptomatic congenital CMV infection, and there was no difference in clinical characteristics and newborn hearing screening results between infants with and without congenital CMV infection at birth.Congenital CMV infection prevalence was lower and the clinical manifestations were milder in this relatively developed region of China compared to populations from other countries with similarly high maternal seroprevalence. Follow-up on children with congenital CMV infection will clarify the burden of disabilities from congenital CMV infection in China.

Wang CH, Yang CY, Lien R, Chu SM, Hsu JF, Fu RH, Chiang MC.
Prevalence and independent risk factors for hearing impairment among very low birth weight infants.
Int J Pediatr Otorhinolaryngol. 2017 Feb;93:123-127. doi:10.1016/j.ijporl.2016.12.029. Epub 2016 Dec 27.

Background: Although we've made big strides in perinatal and neonatal care, auditory handicap remains a serious complication in those who were born very premature.
Objectives: The aim was to determine the prevalence and analyze possible risk factors of hearing impairment in very-low-birth-weight (VLBW) infants.
Materials and Methods: This was a retrospective study by reviewing medical records of all VLBW infants (BW ≤ 1500 g) admitted to NICU of Chang Gung Children's Hospital over 2 years period from Jan. 2010 to 2011. Brainstem auditory evoked potentials (BAEP) hearing screening was performed at 3 months postnatal corrective age and repeated if failed the 1st time, then refer to ENT doctor if BAEP confirmed abnormal. All VLBW infants examined for hearing impairment were included and data were retrieved retrospectively and analyzed for neonatal risk factors using logistic regression.
Results: Over the period, 309 VLBW infants were screened. Prevalence of uni- or bilateral hearing impairment was 3.9% (12/309; 95% CI 2.6-4.1). The mean corrective age on diagnosed of hearing impairment was 2.9 ± 1.1 (range 1-5) months. Mean gestational age was 27.9 weeks (SD 1.4) and mean birth weight was 1028 g (SD 180). By univariant analysis for hearing impairment, severe birth asphyxia, craniofacial anomalies, ventilator dependence, patent ductus arteriosus ligation, and use of postnatal ototoxins yielded good prediction of hearing impairment in this population. However, using multivariate analysis revealed that the only independent risk factors for hearing impairment were ototoxins (OR: 3.62; CI: 1.67-7.82), PDA ligation (OR: 4.96; CI: 2.34-10.52), craniofacial anomalies (OR: 3.42; CI: 1.70-6.88)and assisted prolonged use of oxygen at gestational age of >36 weeks (OR: 5.94; CI: 2.61-13.54).
Conclusion: The incidence of hearing impairment among VLBW infants was 3.9%. Prolonged supplemental oxygen use is a marker for predicting hearing impairment; this requires detailed analysis of the pathophysiologic features, to reduce the prevalence of hearing impairment.

Winiger AM, Alexander JM, Diefendorf AO.
Minimal Hearing Loss: From a Failure-Based Approach to Evidence-Based Practice.
Am J Audiol. 2016 Sep 1;25(3):232-45. doi: 10.1044/2016_AJA-15-0060.

Purpose: A representative sample of the literature on minimal hearing loss (MHL) was reviewed to provide evidence of challenges faced by children with MHL and to establish the need for evidence-based options for early intervention.
Method: Research articles published from 1950 to 2013 were searched in the Medline database using the keywords minimal hearing loss, unilateral hearing loss, and mild hearing loss. References cited in retrieved articles were also reviewed.
Results: In total, 69 articles contained relevant information about pediatric outcomes and/or intervention for unilateral hearing loss, 50 for mild hearing loss, and 6 for high-frequency hearing loss. Six challenges associated with MHL emerged, and 6 interventions were indicated. Evidence indicates that although some individuals may appear to have no observable speech-language or academic difficulties, others experience considerable difficulties. It also indicates that even though children with MHL may appear to catch up in some areas, difficulties in select domains continue into adulthood.
Conclusions: Evidence indicates significant risks associated with untreated MHL. Evidence also demonstrates the need for early intervention and identifies several appropriate intervention strategies; however, no single protocol is appropriate for all children. Therefore, families should be educated about the impact of MHL and about available interventions so that informed decisions can be made.

Wroblewska-Seniuk K, Greczka G, Dabrowski P, Szyfter W, Mazela J.
The results of newborn hearing screening by means of transient otoacoustic emissions - has anything changed over 10 years?
Int J Pediatr Otorhinolaryngol. 2017 May;96:4-10. doi: 10.1016/j.ijporl.2017.02.021. Epub 2017 Feb 21.

Objectives: Universal newborn hearing screening (UNHS) has become the standard of care in many countries. The aim of this study was to evaluate the results of UNHS after ten years of the program in Poland and to compare them with the results of 2003.

Methods: In the study, we analyze the results of UNHS in the University Hospital in Poznan, Poland. Between 01.01.2013 and 31.12.2013, 6827 children were examined by means of otoacoustic emissions.
Results: Risk factors (RF) were identified in 772 (11.3%) newborns, which is significantly less than 10 years ago ($p < 0.05$). The most frequent RF were: ototoxic medications, treatment in neonatal intensive care unit (NICU) and prematurity < 33 weeks of gestation. In 2003, the most frequent were ototoxic medications and prematurity, less frequent was treatment in NICU and more common was low Apgar score. In 51 (6.6%) newborns with RF, the result of OAE was positive either unilaterally or bilaterally. In infants without RF the result was positive unilaterally in 22 (0.4%) and bilaterally in 14 (0.2%) patients. These results are significantly lower than in our former study. The relative risk of positive result was the highest in infants with complex congenital anomalies (RR = 44.99), craniofacial anomalies (RR = 17.46) and mechanical ventilation for > 5 days (RR = 10.69). In our previous study, the highest RR of positive test results was in infants with family history, congenital malformations and low Apgar score. We found that most predictive as to the final diagnosis was bilaterally positive OAE test. In most patients, the second check confirmed the diagnosis, independently of RF. The number of false positive tests at the 1st level of screening is significantly lower now than 10 years ago, probably due to better staff training.
Conclusions: Long term monitoring and the appropriate management of hearing deficit in children is essential. UNHS seems to be the most efficient way of finding children who require treatment of hearing impairment. The prevalence of most risk factors of hearing deficit has significantly changed over the years. The number of false positive results has significantly decreased over the years thanks to better staff training.

Wroblewska-Seniuk KE, Dabrowski P, Szyfter W, Mazela J.
Universal newborn hearing screening: methods and results, obstacles, and benefits.
Pediatr Res. 2017 Mar;81(3):415-422. doi: 10.1038/pr.2016.250. Epub 2016 Nov 18.
The incidence of sensorineural hearing loss ranges from 1 to 3 per 1,000 live births in term healthy neonates, and 2-4 per 100 in high-risk infants, a 10-fold increase. Early identification and intervention with hearing augmentation within 6 mo yields optimal effect. If undetected and without treatment, significant hearing impairment may negatively impact speech development and lead to disorders in psychological and mental behaviors. Hearing screening programs in newborns enable detection of hearing impairment in the first days after birth. Programs to identify hearing deficit have significantly improved over the two decades, and their implementation continues to grow throughout the world. Initially based on risk factors, these programs identified only 50-75% of infants with hearing loss. Current recommendations are to conduct universal hearing screening in all infants. Techniques used primarily include automated auditory brainstem responses and otoacoustic emissions that provide noninvasive recordings of physiologic auditory activity and are easily performed in neonates and infants. The aim of this review is to present the objectives, benefits, and results of newborn hearing screening programs including the pros and cons of universal vs. selective screening. A brief history and the anticipated future development of these programs will also be discussed.

Wu CC, Tsai CH, Hung CC, Lin YH, Lin YH, Huang FL, Tsao PN, Su YN, Lee YL, Hsieh WS, Hsu CJ.
Newborn genetic screening for hearing impairment: a population-based longitudinal study.
Genet Med. 2017 Jan;19(1):6-12. doi: 10.1038/gim.2016.66. Epub 2016 Jun 16.
Purpose: The feasibility of genetic screening for deafness-causing mutations in newborns has been reported in several studies. The aim of this study was to investigate the long-term results in those who screened positive for deafness mutations; these results are crucial to determine the cost-effectiveness to justify population-wide genetic screening.
Methods: We performed simultaneous hearing screening and genetic screening targeting four common deafness mutations (p.V37I and c.235delC of GJB2, c.919-2A>G of SLC26A4, and the mitochondrial m.1555A>G) in 5173 newborns at a tertiary hospital between 2009 and 2015. Serial audiometric results up to 6 years old were then analyzed in children with conclusive genotypes.
Results: Newborn genetic screening identified 82 (1.6%) babies with conclusive genotypes, comprising 62 (1.2%) with GJB2 p.V37I/p.V37I, 16 (0.3%) with GJB2 p.V37I/c.235delC, and 4 (0.1%) with m.1555A>G. Of these, 46 (56.1%) passed hearing screening at birth. Long-term follow-up demonstrated progressive hearing loss in children with the GJB2 p.V37I/p.V37I and p.V37I/c.235delC genotypes; this hearing loss deteriorated by approximately 1 decibel hearing level (dBHL) per year.
Conclusion: We delineated the longitudinal auditory features of the highly prevalent GJB2 p.V37I mutation on a general population basis and confirmed the utility of newborn genetic screening in identifying infants with late-onset or progressive hearing impairment undetectable by newborn hearing screening.

Wu GT, Devine C, Xu A, Geelan-Hansen K, Anne S.
Is routine audiometric testing necessary for children with isolated preauricular lesions?
Int J Pediatr Otorhinolaryngol. 2017 Feb;93:68-70. doi: 10.1016/j.ijporl.2016.12.032. Epub 2016 Dec 27.
Introduction: Preauricular lesions, including tags, pits, sinuses, and cysts are commonly seen. Some studies have shown increased incidence of hearing loss in these patients but other studies have failed to corroborate this finding. The purpose of this study is to evaluate the incidence of hearing loss in patients with isolated preauricular lesions.

Methods: Retrospective chart review of all pediatric otolaryngology patients seen at a tertiary academic center between 2008 and 2014. All patients with the diagnosis code of 744.1 or 701.9 (preauricular skin tag) or 744.46, 744.47, or 744.89 (preauricular pit/fistula/cyst) were included in this study. Medical records were reviewed for clinical, demographic, and audiologic data.
Results: Ninety-nine patients, 46 males, 53 females, with preauricular lesions were identified. Twelve were found to have abnormal hearing. Five patients had conductive hearing loss due to underlying Eustachian tube dysfunction. Four patients had sensorineural hearing loss; three of these patients had an enlarged vestibular aqueduct and one patient did not have an identified cause. Three patients had sound field testing or abnormal otoacoustic emissions that suggested hearing loss with no further follow up.
Conclusion: Children with isolated preauricular lesions with no history of otologic surgery or risk factors for hearing loss may not need audiologic evaluation outside of regular hearing screening. However, there does appear to be a higher association with Eustachian tube dysfunction in these children. Further studies will need to be done to determine whether or not there is an embryological correlation for this finding.

Yamaguchi A, Oh-Ishi T, Arai T, Sakata H, Adachi N, Asanuma S, Oguma E, Kimoto H, Matsumoto J, Fujita H, Uesato T, Fujita J, Shirato K, Ohno H, Kizaki T.
Screening for seemingly healthy newborns with congenital cytomegalovirus infection by quantitative real-time polymerase chain reaction using newborn urine: an observational study.
BMJ Open. 2017 Jan 20;7(1):e013810. doi: 10.1136/bmjopen-2016-013810.

Objective: Approximately 8-10% of newborns with asymptomatic congenital cytomegalovirus (cCMV) infection develop sensorineural hearing loss (SNHL). However, the relationship between CMV load, SNHL and central nervous system (CNS) damage in cCMV infection remains unclear. This study aimed to examine the relationship between urinary CMV load, SNHL and CNS damage in newborns with cCMV infection.
Study Design: The study included 23 368 newborns from two maternity hospitals in Saitama Prefecture, Japan. Urine screening for cCMV infection (quantitative real-time PCR) and newborn hearing screening (automated auditory brainstem response (AABR) testing) were conducted within 5 days of birth to examine the incidence of cCMV infection and SNHL, respectively. CNS damage was assessed by MRI of cCMV-infected newborns.
Results: The incidence of cCMV infection was 60/23 368 (0.257%; 95% CI 0.192% to 0.322%). The geometric mean urinary CMV DNA copy number in newborns with cCMV was 1.79×10^6 copies/mL (95% CI 7.97×10^5 to 4.02×10^6). AABR testing revealed abnormalities in 171 of the 22 229 (0.769%) newborns whose parents approved hearing screening. Of these 171 newborns, 22 had SNHL (12.9%), and 5 of these 22 were infected with cCMV (22.7%). Newborns with both cCMV and SNHL had a higher urinary CMV DNA copy number than newborns with cCMV without SNHL (p=0.036). MRI revealed CNS damage, including white matter abnormalities, in 83.0% of newborns with cCMV. Moreover, newborns with CNS damage had a significantly greater urinary CMV load than newborns without CNS damage (p=0.013).
Conclusions: We determined the incidence of cCMV infection and urinary CMV DNA copy number in seemingly healthy newborns from two hospitals in Saitama Prefecture. SNHL and CNS damage were associated with urinary CMV DNA copy number. Quantification of urinary CMV load may effectively predict the incidence of late-onset SNHL and neurodevelopmental disorders.
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Yang HC, Sung CM, Shin DJ, Cho YB, Jang CH, Cho HH.
Newborn hearing screening in prematurity: fate of screening failures and auditory maturation.
Clin Otolaryngol. 2017 Jun;42(3):661-667. DOI:10.1111/coa.12794. 12794. Epub 2016 Dec 7.

Objectives: The purpose of this study was to identify delayed auditory maturation and the fate of premature infants who failed the newborn hearing screening (NHS) in neonatal intensive care unit.
Materials and Methods: A total of 1375 neonates underwent NHS using the transient evoked otoacoustic emission (TEOAE) in a tertiary hospital between 2007 and 2010 according to the Joint Committee on Infant Hearing guidelines. In addition, a structured telephone survey was given to caregivers of infants who were lost to follow-up NHS. Auditory steady-state response (ASSR) threshold and the threshold change in diagnostic test failures were analysed.
Result: Among the 1375 NICU babies, 344 (25.0%) babies, 111 (9.7%) babies and 64 (4.6%) babies failed to pass the first TEOAE, second TEOAE and diagnostic ASSR, respectively. However, at the age of about 5 years, 12 (0.9%) infants showed permanent hearing loss (PHL). The ASSR threshold improved from 69.0 ± 19.7 dB to 52.9 ± 21.6 dB in <4 months ($P < 0.001$). Premature infants of <29 weeks of gestational age at birth showed higher referral ($P = 0.003$) rate at the first OAE test compared to the others, and the difference continued until the last follow-up. The odds ratio for the initial ASSR threshold >67.5 dB for PHL was 9.00 (95% confidence interval, 1.7-46.7).
Conclusion: Most of first TEOAE screening failures (91.3%) showed normal hearing and speech development. Hearing levels in premature infants can improve over time, particularly in neonates with initial ASSR threshold <67.5 dB.

Yang SM, Liu Y, Liu C, Yin AH, Wu YF, Zheng XE, Yang HM, Yang J.

Hearing-loss-associated gene detection in neonatal intensive care unit.

J Matern Fetal Neonatal Med. 2017 Mar 27:1-5. doi: 10.1080/14767058.2017.1282454. [Epub ahead of print]

Objective: To investigate the frequency and mutation spectrum of hearing loss-associated gene mutation in Neonatal Intensive Care Unit (NICU).

Methods: Neonates (n=2305) admitted to NICU were enrolled in this study. Nine prominent hearing loss-associated genes, GJB2 (35 del G, 176 del 16,235 del C, 299 del AT), GJB3 (538 C>T), SLC26A4 (IVS7-2A>G, 2168 A>G) and mtDNA 12S rRNA(1555 A>G, 1494 C>T), were detected.

Result: There were 73 cases hearing-loss-associated gene mutation among 2305 cases, the mutation frequency was 3.1%, with 40 cases GJB2 (235del C) mutation (54.8%), 6 cases GJB2 (299 del AT) mutation (8.2%), 21 cases SLC26A4 (IVS 7-2 A>G) mutation (28.7%), 4 cases SLC26A4 (2168 A>G) mutation (5.5%), 2 cases of GJB2 (235del C) combined SLC26A4 (IVS 7-2 A>G, 2168 A>G) mutation (2.8%). Among 73 gene mutation cases, preterm neonates presented in 18 cases, accounting for 24.7% (18/73); hyperbilirubinemia in 13 cases, accounting for 17.8% (13/73); Torch Syndrome in 15 cases, with 12 cases CMV, 2 cases rubella, 1 case toxoplasm, respectively, totally accounting for 20.54% (15/73); neonatal pneumonia in 12 cases, accounting for 16.4% (12/73); birth asphyxia in 5 cases, accounting for 6.9% (5/73); sepsis in 5 cases, accounting for 6.9% (5/73); others in 5 cases, accounting for 6.8% (5/73) .

Conclusion: The frequency of hearing loss-associated gene mutation was higher in NICU.There were hearing loss-associated gene mutations in the NICU, suggesting this mutation may complicate with perinatal high-risk factors.

Zeitlin W, Auerbach C, Mason SE, Spivak LG, Reiter B.

Factors Related to Not Following Up with Recommended Testing in the Diagnosis of Newborn Hearing Loss.

Health Soc Work. 2017 Feb 1;42(1):24-31. doi: 10.1093/hsw/hlw061.

Children's hearing is a public health concern, and universal newborn hearing screenings are the first step in detecting and treating congenital hearing loss. Despite the high rate of participation in such programs, loss to follow-up (LTF) with additional recommended diagnosis and treatment has been a persistent problem. The current research seeks to expand the knowledge base at the point of diagnosis, where there is a large drop-off in parents following through with recommended care. This research was organized around the following question: What biopsychosocial factors are associated with LTF between screenings and diagnostic evaluations? A prospective quantitative longitudinal study tracked 203 families whose newborns were referred for additional testing at discharge from the hospital after birth. Binary logistic regression was used to determine what constellation of factors best predicted LTF. Psychosocial factors related to being lost to follow-up at diagnosis included race and ethnicity and access to health care professionals, with African American babies being most at risk for LTF; however, the impact of race and ethnicity declined when parents believed they had more health care professionals with whom to consult.