

## Others' Publications About EHDI: April through October, 2018

The *Journal of Early Hearing Detection and Intervention (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, and quality improvement that contribute to improving Early Hearing Detection and Intervention (EHDI) systems.

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

EHDI continues to be a global phenomenon with dozens of articles reporting on the status and progress of newborn hearing screening programs—from a systematic analysis of how Poland's national universal newborn hearing screening program has evolved over the last 15 years to reports of emerging newborn hearing screening programs in Iran, Malaysia, and Botswana, among others. These reports emphasize the importance of tailoring screening and follow-up programs to local conditions and practices. There continues to be a major focus on targeted and universal screening for congenital cytomegalovirus and genetic mutations associated with hearing loss. A number of articles reported on successful efforts to screen and identify hearing loss in young children, recognizing that there are as many or more children who acquire hearing loss after the neonatal period as there are children with congenital hearing loss. The broadening of screening and identification programs to other conditions that are associated with hearing loss and to young children beyond the newborn period has important implications for how newborn hearing screening programs are best implemented and managed. Staying current with EHDI's processes and accomplishments requires staying informed about what is happening in these closely related areas

Below are some of the interesting findings from recently reported research studies from around the world.

- Lantos et al., in a health disparities study conducted in the United States, found that urban, low-income neighborhoods had a higher prevalence of infant hearing loss compared with more affluent surrounding communities, particularly among minorities.
- ElAlfy et al., in a study conducted in Egypt, found that iron deficiency anemia during late pregnancy adversely affected the newborn's hearing status. They recommended antenatal screening of pregnant mothers to improve fetal iron status and prevent abnormal auditory maturation for the newborn.
- Rahimi et al., in a five-year study conducted in Iran with 4,729 children, reported that doing hearing screening during a neonatal thyroid screening program done during the third to fifth day after birth, significantly improved hearing screening outcomes. They concluded that this approach could be used as an alternative to hospital-based newborn hearing screening in many countries
- Banda et al., demonstrated the importance of screening children for hearing loss after the newborn period by reporting on the high incidence of hearing loss among children 10 years of age and younger who were referred to a public audiology clinic in Botswana.
- Hao et al., based on a cohort of 142,417 neonates in China who were screened for common genetic mutations associated with hearing loss, concluded that genetic screening for hearing loss was practical in a large-scale community setting.
- Cejas et al., reported on a longitudinal study of 147 children with cochlear implants and 75 typically hearing peers. They found that children using cochlear implants perform similarly to hearing peers on measures of intelligence, but those with severe comorbidities are at-risk for cognitive deficits.
- Fowler and Boppana provide an excellent overview of congenital cytomegalovirus, how it is related to childhood hearing loss, and why it should be carefully considered as a part of early hearing detections and intervention systems.

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

Adachi K, Xu J, Ank B, Watts DH, Camarca M, Mofenson LM, Pilotto JH, Joao E, Gray G, Theron G, Santos B, Fonseca R, Kreitchmann R, Pinto J, Mussi-Pinhata MM, Machado DM, Ceriotta M, Morgado MG, Bryson YJ, Veloso VG, Grinsztejn B, Mirochnick M, Moya J, Nielsen-Saines K; MPH for the NICHD HPTN 040 Study Team.

[Congenital Cytomegalovirus and HIV Perinatal Transmission. \*Pediatr Infect Dis J\*.](#)

2018 Oct;37(10):1016-1021. doi: 10.1097/INF.0000000000001975.

**BACKGROUND:** Congenital cytomegalovirus (CMV) infection (cCMV) is an important cause of hearing loss and cognitive impairment. Prior studies suggest that HIV-exposed children are at higher risk of acquiring cCMV. We assessed the presence, magnitude and risk factors associated with cCMV among infants born to HIV-infected women, who were not receiving antiretrovirals during pregnancy.

**METHODS:** cCMV and urinary CMV load were determined in a cohort of infants born to HIV-infected women not receiving antiretrovirals during pregnancy. Neonatal urines obtained at birth were tested for CMV DNA by qualitative and reflex quantitative real-time polymerase chain reaction.

**RESULTS:** Urine specimens were available for 992 (58.9%) of 1684 infants; 64 (6.5%) were CMV-positive. Mean CMV load (VL) was 470,276 copies/ml (range: < 200-2,000,000 copies/ml). Among 89 HIV-infected infants, 16 (18%) had cCMV versus 42 (4.9%) of 858 HIV-exposed, uninfected infants ( $P < 0.0001$ ). cCMV was present in 23.2% of infants with in utero and 9.1% infants with intrapartum HIV infection ( $P < 0.0001$ ). Rates of cCMV among HIV-infected infants were 4-fold greater (adjusted OR, 4.4; 95% CI: 2.3-8.2) and 6-fold greater among HIV in utero-infected infants (adjusted OR, 6; 95% CI: 3-12.1) compared with HIV-exposed, uninfected infants. cCMV was not associated with mode of delivery, gestational age, Apgar scores, 6-month infant mortality, maternal age, race/ethnicity, HIV viral load or CD4 count. Primary cCMV risk factors included infant HIV-infection, particularly in utero infection.

**CONCLUSION:** High rates of cCMV with high urinary CMV VL were observed in HIV-exposed infants. In utero HIV infection appears to be a major risk factor for cCMV in infants whose mothers have not received combination antiretroviral therapy in pregnancy.

Adegbiyi WA, Olajide GT, Olatoke F, Olajuyin AO, Olubi O, Ali A, Eletta PA, Aluko AA.

[Preschool Children Hearing Impairment: Prevalence, Diagnosis and Management in a Developing Country. \*Int Tinnitus J\*.](#)

2018 Jun 1;22(1):60-65. doi: 10.5935/0946-5448.20180010.

**OBJECTIVES:** Preschool children hearing impairment is a common otologic diseases worldwide. The burdens of this preventable condition can be reduced in developing country. This study was carried out to determine the prevalence, diagnosis and management of hearing impairment among children less than 5 years seen in our center.

**METHOD:** This was a prospective hospital based study of preschool children with complaints of hearing impairment in Ekiti state university teaching hospital, Nigeria. This study was conducted over a period of two years (February 2016 to January 2018). Data was obtained from consented patients by using pretested interviewers assisted questionnaire. Data obtained was collated and analyzed by using SPSS software version 18.0. Descriptive statistics was used to present and expressed the data as simple tables and charts.

**RESULTS:** The prevalence of hearing impairment in this study was 5.9% with peak value of 37.6% at preschool age 3 years. There were 42.6% males and 57.4% females. Majority (51.5%) of the patients were urban dwellers. The aetiologic factors of hearing impairment include; 15.8% neonatal jaundice, 13.9% febrile illness, 12.9% otitis media and 11.9% birth asphyxia. Pre-lingual presentation was commonest in 57.4%. Common clinical features includes yet to speak, restlessness/stubborn and not responding to command in 60.4%, 48.5% and 46.5% respectively. Commonest type of hearing impairment was sensorineural hearing loss is 61.4%. Type A tympanometry (normal) was the commonest findings in 82.2%.

**CONCLUSION:** Preschool children hearing impairment with late presentation is a common otologic diseases burden to parent and otorhinolaryngologist in our center. The common causes were preventable with resulting permanent auditory damage.

Ahmed S, Sheraz S, Malik SA, Ahmed NR, Malik SA, Farooq S, Raheem A, Basheer F, Nayyar ZA; Fazal-e-Malik.

[Frequency Of Congenital Hearing Loss In Neonates. \*J Ayub Med Coll Abbottabad\*.](#)

2018 Apr-Jun;30(2):234-236.

**BACKGROUND:** Congenital deafness is commonest birth defect and it affects 2-4 neonates among 1000 live births. Detection and intervention especially before 6 months of age prevents severe linguistic, educational and psychosocial repercussions and helps the deaf child in the development of normal speech and language. Children who are identified after 6 months of age experience great difficulties in attaining speech and language..

**METHODS:** To find out the frequency of hearing loss in neonates, a hospital based observational study was conducted in Combined Military Hospital Abbottabad from June-to December 2014. One thousand new-borns selected by consecutive sampling within a specified period of time were subjected to Otoacoustic Emission (OAE) testing. Brain Evoked Response Audiometry (BERA) evaluation was performed in all those who failed OAE testing to confirm the hearing loss. Children born with microtia, meatal stenosis, cleft palate, craniofacial abnormalities and syndromic illnesses were excluded from the study.

**RESULTS:** Of 1000 newborns screened, 465 were males and 535 were females whereas 632 (63.2%) were delivered through C-section and 368 (36.8%) were born via SVD. Four hundred and ninety-one (49%) babies had a positive history of consanguinity among the parents. Out of 1000 infants 13 were having hearing loss which was later on confirmed on BERA evaluation. Among them 7 were males and 6 females, 9 (69%) were born through SVD and 4 (31%) through C-section and 8 (61.5%) new-borns had a positive history of consanguinity among their parents. In all these 13 patients only 2 (15%) patients had profound while the remaining 11 (85%) had moderate to severe hearing loss.

**CONCLUSIONS:** Frequency of hearing loss in neonates is much higher in our study (13 per 1000) as compared to other parts of the world and demands that more studies should be undertaken on this subject to confirm this.

Almosnino G, Anne S, Schwartz SR.

[Use of Neural Response Telemetry for Pediatric Cochlear Implants: Current Practice. Ann Otol Rhinol Laryngol.](#)

2018 Jun;127(6):367-372. doi: 10.1177/0003489418767692. Epub 2018 Apr 12.

**OBJECTIVES:** Evaluate usage trends of neural response telemetry (NRT) in cochlear implant centers across the nation and assess reported benefits of intraoperative NRT for pediatric cochlear implant recipients.

**STUDY DESIGN:** Survey.

**STUDY PARTICIPANTS:** All US cochlear implant centers (n = 110).

**METHODS:** A 15-question multiple-choice survey was distributed electronically to all centers. The survey captured demographic information of all centers, practice patterns surrounding the use of NRT, and the extent to which intraoperative NRT is of benefit.

**RESULTS:** Thirty-two invited participants (29%) completed the survey. A majority of participants reported practicing in an academic center (66%), followed by a hospital setting (19%) and private practice (16%). Seventy-two percent of survey participants reported using NRT for pediatric cochlear implant recipients. Sixty-three percent felt it improved the ability to program at initial activation, and 50% of participants felt that NRT improves satisfaction at initial activation.

**CONCLUSION:** This study suggests that a majority of surgeons use intraoperative NRT for pediatric cochlear implantation as an additional measure to ensure appropriate electrode placement and improve device activation. Larger studies are needed to better establish the relationship between intraoperative NRT and postoperative outcomes and justify the additional costs associated with intraoperative NRT.

Banda FM, Powis KM, Mokoka AB, Mmapetla M, Westmoreland KD, David T, Steenhoff AP.

[Hearing Impairment Among Children Referred to a Public Audiology Clinic in Gaborone, Botswana. Glob Pediatr Health.](#)

2018 Apr 20;5:2333794X18770079. doi: 10.1177/2333794X18770079. eCollection 2018.

*Objective.* To describe and quantify hearing impairment among children referred to the audiology clinic in Princess Marina Hospital, a public referral hospital in Botswana. *Methods.* In a retrospective case series, we reviewed medical records of children aged 10 years and younger whose hearing was assessed between January 2006 and December 2015 at the audiology clinic of Princess Marina Hospital in Gaborone, Botswana. *Results.* Of 622 children, 50% were male, and median age was 6.7 years (interquartile range = 5.0-8.3). Hearing impairment was diagnosed in 32% of clinic attendees, comprising sensorineural (23%), conductive (25%), and mixed (11%) hearing loss, while 41% of children with diagnosed hearing impairment did not have a classification type. Hearing impairment was mild in 22.9%, moderate in 22.4%, severe in 19.4%, profound in 16.9%, and of undocumented severity in 18.4%. Children younger than 5 years were 2.7 times (95% confidence interval = 1.29-5.49;  $P = .008$ ) more likely to be diagnosed with sensorineural hearing impairment compared with those older than 5 years. By contrast, children older than 5 years were 9.6 times (95% confidence interval = 2.22-41.0;  $P = .002$ ) more likely to be diagnosed with conductive hearing loss compared with those under 5 years. *Conclusion.* Hearing impairment was common among children referred to this audiology clinic in Botswana. Of those with hearing impairment, more than a third had moderate or severe deficits, suggesting that referrals for hearing assessments are not occurring early enough. Hearing awareness programs individually tailored to parents, educators, and health care workers are needed. Neonatal and school hearing screening programs would also be beneficial.

Bartlett AW, Hall BM, Palasanthiran P, McMullan B, Shand AW, Rawlinson WD.

[Recognition, treatment, and sequelae of congenital cytomegalovirus in Australia: An observational study. J Clin Virol.](#)

2018 Sep 27;108:121-125. doi: 10.1016/j.jcv.2018.09.017. [Epub ahead of print]

**BACKGROUND AND OBJECTIVES:** Australian national surveillance data was used to assess recognition, sequelae, and antiviral therapy for congenital cytomegalovirus (CMV) cases.

**STUDY DESIGN:** Data from congenital CMV cases reported through the Australian Paediatric Surveillance Unit born January 1999 to December 2016 were described and Chi-square tests used to characterise trends and associations in case reporting, maternal CMV serology testing, and antiviral therapy. Descriptive analyses for hearing loss and developmental delay were reported for cases born  $\geq 2004$ , following introduction of universal neonatal hearing screening.

**RESULTS:** There were 302 congenital CMV cases (214 symptomatic, 88 asymptomatic). Congenital CMV was suspected in 70.6% by 30 days of age, with no differences across birth cohorts. Maternal CMV serology testing was associated with maternal illness during pregnancy but not birth cohort. There was increasing antiviral use for symptomatic cases, being used in 14% born 1999-2004, 19.6% born 2005-2010, and 44.4% born 2011-2016 ( $p < 0.001$ ). For those born  $\geq 2004$ , hearing loss was reported in 42.1% of symptomatic and 26.6% of asymptomatic cases; while developmental delay was reported in 16.9% of symptomatic and 1.3% of asymptomatic cases.

**CONCLUSION:** There appears to be under-reporting and under-recognition of congenital CMV despite increasing use of antiviral therapy. Universal newborn CMV screening should be considered to facilitate follow-up of affected children and targeted linkage into hearing and developmental services, and to provide population-level infant CMV epidemiology to support research and evaluation of antiviral and adjunctive therapies.

**Bhindi A, Carpineta L, Qassabi B, Waissbluth S, Ywakim R, Manoukian JJ, Nguyen LHP.**

**[Hearing loss in pediatric temporal bone fractures: Evaluating two radiographic classification systems as prognosticators.](#) *Int J Pediatr Otorhinolaryngol.* 2018 Jun;109:158-163. doi: 10.1016/j.ijporl.2018.04.005. Epub 2018 Apr 12.**

**INTRODUCTION:** Temporal bone fractures (TBF) are traditionally classified by their angle of fracture relative to the petrous ridge, and more recently by whether or not they violate the otic-capsule. This study compared rates of hearing loss (HL) and signs of otologic dysfunction among fracture types of both classification systems, within the pediatric population.

**METHODS:** Pediatric patients were retrospectively characterized from a previously identified cohort of TBF patients, diagnosed from 2000 to 2014. CT scans were reviewed and TBFs were classified first as longitudinal (L), transverse (T) or mixed (M), and then as otic-capsule sparing (OCS) or otic-capsule violating (OCV). Medical records were reviewed, and rates of HL and presenting signs were compared among L, T and M fractures, and OCS and OCV fractures.

**RESULTS:** Forty-three patients with 47 TBFs met the inclusion criteria. Eighteen, 4 and 25 TBFs were classified as L, T and M fractures, respectively. Thirty-three and 9 were classified as OCS, and OCV, respectively. Among 24 cases of HL: 20, 3, and 1 were conductive HL (CHL), sensorineural HL (SNHL) and mixed HL, respectively. Two cases of SNHL were found among OCV fractures, with none in OCS fractures (estimated difference 0.22; 95% confidence interval 0.01-0.60). Similar rates of CHL were found across L, T and M fractures (range 36-50%), and across OCV and OCS fractures (range 42-44%). Hemotympanum was the most common presenting sign, found in 68% of TBFs and 80% of CHL cases. There were no significant differences in the incidence of signs or symptoms between fracture types.

**CONCLUSIONS:** In our cohort, both the traditional and otic-capsule radiographic classification systems failed to predict the incidence of CHL and other otologic signs in the pediatric population. Though OCV fractures conferred an increased risk for developing SNHL, we found a lower incidence than anticipated given violation to the bony labyrinth.

**Boskabadi H, Zakerihamidi M, Moradi A, Bakhshae M.**

**[Risk Factors for Sensorineural Hearing Loss in Neonatal Hyperbilirubinemia.](#) *Iran J Otorhinolaryngol.***

2018 Jul;30(99):195-202.

**Introduction:**Hyperbilirubinemia is a common neonatal problem with toxic effects on the nervous system that can cause hearing impairment. This study was conducted to assess the risk factors for sensorineural hearing loss and other coexisting problems in icteric infants.

**Materials and Methods:** In a case-control study, 200 term infants with bilirubin levels higher than 20 mg/dl admitted to the neonatal intensive care unit of Ghaem Hospital, Mashhad during 2007-2015 were investigated. Profiles of infants with hearing impairment (n=60) were compared with those of icteric newborns with normal hearing (140 newborns) as the control group. After confirming the clinical diagnosis of jaundice by laboratory findings, a validated questionnaire containing mother and infant profiles were used for data collection. The auditory brainstem response test was used for assessment of infant hearing status after discharge.

**Results:** Sensorineural hearing loss among infants with severe hyperbilirubinemia was found to be 4.8%. Serum total bilirubin (P=0.001), creatinine levels (P=0.002), direct Coombs test results (P=0.001), etiology (P=0.000) and treatment for jaundice (P=0.000), eye movement disorders (P=0.001), opisthotonos (P=0.001), and microcephaly (P=0.001) were found to be significantly different between the two groups (P<0.005). The prognostic predictability of sensorineural hearing loss based on total bilirubin level was found to be 82%.

**Conclusion:** Hearing impairment occurs about 10-50 times more frequently in neonates with severe jaundice. Total bilirubin level has the highest predictability for infant hearing status. Blood group and Rhesus (Rh) incompatibilities between mother and child and G6PD deficiency are important known causes for hearing impairment due to jaundice.

**Chakrabarti S.**

**[State of deaf children in West Bengal, India: What can be done to improve outcome.](#) *Int J Pediatr Otorhinolaryngol.***

2018 Jul;110:37-42. doi: 10.1016/j.ijporl.2018.04.025. Epub 2018 Apr 27.

**OBJECTIVE:** Prelingual deafness in children demands urgent action as best outcome is dependent on earliest possible diagnosis and intervention. Objective of this study was to determine age of suspicion, diagnosis, intervention, and outcome in a representative group of deaf children in West Bengal, India, and suggest ways of improving these parameters.

**METHODS:** In this cross-sectional study, ages of suspicion, diagnosis, intervention and outcome of 303 randomly selected deaf children were elicited from a cohort of 1316 children with deafness identified in an earlier study.

**RESULTS:** Median ages of suspicion, diagnosis and amplification were 18, 72 and 84 months respectively. Age of suspicion was significantly related to parental education (p < 0.05); age of diagnosis to parental education and socio-economic status (p < 0.001) and children's geographic location (p < 0.01). Following diagnosis, 86% of children received hearing aids but only 6% used their aids consistently; 86% were non-verbal, 12% could communicate with a mixture of speech and gesture and only 2% with speech alone.

**CONCLUSION:** Current situation of deaf children in West Bengal, and evidence indicates, in much of India, is insupportable. However, widely diverse socio-economic conditions and scarcity of public health infrastructure preclude one solution of the problem for the whole country. In absence of the ideal universal newborn hearing screening, rigorously monitored and costed pilot programs of different models of early detection and intervention using newborn hearing screening, targeted screening and trial of calibrated noisemakers by primary care workers should be tried to see which works best where, so that successful programs can be scaled up over time.



Chen JY, Yang J.

[International consensus \(ICON\) on audiological assessment of hearing loss in children. Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi.](#)

2018 Jun;32(12):886-890. doi: 10.13201/j.issn.1001-1781.2018.12.002.[Article in Chinese]

**Summary** The prevalence of hearing loss in newborns and infants is estimated between 1 to 3.47 cases per 1000 live births. Neonatal screening for hearing loss and audiological evaluation are becoming more extensively carried out. However, there is no consensus regarding the use of audiometry and other electrophysiological tests in current practices. This article is intended to provide professionals with recommendations about the “best practice” based on consensus opinion of the session’s speakers, and a review of the literature on the efficacy of various assessment options for children with hearing loss.

Cevizci R, Dilci A, Celenk F, Karamert R, Bayazit Y.

[Surgical considerations and safety of cochlear implantation in otitis media with effusion. Auris Nasus Larynx.](#)

2018 Jun;45(3):417-420. doi: 10.1016/j.anl.2017.07.012. Epub 2017 Jul 26.

**OBJECTIVE:** To evaluate the effects of otitis media with effusion on surgical parameters, patient safety, perioperative and postoperative complications.

**METHODS:** Total 890 children who underwent cochlear implantation between 2006 and 2015 were included. The ages ranged from 12 months to 63 months (mean: 32 months). The patients were divided into two groups according to the presence or absence of otitis media with effusion; otitis media with effusion group and non-otitis media group.

**RESULTS:** Of 890 children, 105 had otitis media with effusion prior to surgery. In non-otitis media with group, there were 785 children. The average duration of surgery was 60min (ranged from 28 to 75min) in non-otitis media group, and 90min (ranged from 50 to 135min) in otitis media with effusion group ( $p<0.05$ ). Granulation tissue and edematous middle ear and mastoid mucosa were observed in all cases of otitis media with effusion during the surgery. There was no significant difference between the complications of groups with or without otitis media with effusion ( $p>0.05$ ). In 5 of 105 patients, there was a ventilation tube inserted before cochlear implantation, which did not change the outcome of implantation.

**CONCLUSION:** There is no need for surgical treatment for otitis media with effusion before implantation since otitis media with effusion does not increase the risks associated with cochlear implantation. Operation duration is longer in the presence of otitis media with effusion. However, otitis media with effusion leads to intraoperative difficulties like longer operation duration, bleeding, visualization of the round window membrane, cleansing the middle ear granulations as well as mastoid and petrous air cells.

Chiong CM, Reyes-Quintos MRT, Yarza TK, Tobias-Grasso CAM, Acharya A, Leal SM, Mohlke KL, Mayol NL, Cutionco-de la Paz EM, Santos-Cortez RLP.

[The SLC26A4 c.706C>G \(p.Leu236Val\) Variant is a Frequent Cause of Hearing Impairment in Filipino Cochlear Implantees. Otol Neurotol.](#)

2018 Sep;39(8):e726-e730. doi: 10.1097/MAO.0000000000001893.

**HYPOTHESIS:** Variants in SLC26A4 are an important cause of congenital hearing impairment in the Philippines.

**BACKGROUND:** Cochlear implantation is a standard rehabilitation option for congenital hearing impairment worldwide, but places a huge cost burden in lower-income countries. The study of risk factors such as genetic variants that may help determine genetic etiology of hearing loss and also predict cochlear implant outcomes is therefore beneficial.

**METHODS:** DNA samples from 29 GJB2-negative Filipino cochlear implantees were Sanger-sequenced for the coding exons of SLC26A4. Exome sequencing was performed to confirm results.

**RESULTS:** Four cochlear implantees with bilaterally enlarged vestibular aqueducts (EVA) were homozygous for the pathogenic SLC26A4 c.706C>G (p.Leu236Val) variant, which has a minor allele frequency of 0.0015 in Filipino controls. In patients with the SLC26A4 variant there was no association between cochlear implant outcome and age at implantation or duration of implant. There was also no association between the occurrence of the SLC26A4 variant and postsurgical audiometric thresholds and parents’ evaluation of aural/oral performance of children (PEACH) scores. On the other hand, the SLC26A4 variant increased presurgical median audiometric thresholds ( $p=0.01$ ), particularly at 500 to 2000Hz.

**CONCLUSION:** The SLC26A4 c.706C>G (p.Leu236Val) variant is a frequent cause of congenital hearing impairment in Filipinos and is associated with bilateral EVA and increased presurgical audiometric thresholds, but does not adversely affect post-implant outcomes.

Chung YS, Park SK.

[Current status of newborn hearing screening in the southeastern region of Korea. Epidemiol Health.](#)

2018 Sep 14. doi: 10.4178/epih.e2018044. [Epub ahead of print]

**Objectives:** To analyze the current status and problems of hearing screening tests for newborns of low income class in the southeastern region of Korea.

**Methods:** This study analyzed the data of the Ministry of Health and Welfare’s project on the early detection of hearing loss in low income class newborns from the southeastern region of Korea (2011 to 2015).

**Results:** The referral rate was 1.33%, 1.69%, and 1.27% in Daegu, Gyeongbuk, and Ulsan, respectively. The confirmation test rate was 36.09%, 23.38%, and 52.94% in Daegu, Gyeongbuk, and Ulsan, respectively. The incidence of hearing loss (adjusted) was 0.41%, 0.62%, and 0.41% in Daegu, Gyeongbuk, and Ulsan, respectively. After confirming the hearing loss, newborns with hearing handicaps were mostly lost to follow up, and rehabilitation, such as hearing aids or cochlear implants, were not used. The screening tests were performed within one month of birth, and the confirmation tests were performed within three months of birth. On the other hand, the groups with the risk factor for hearing loss took more than three months to reach the

date of confirmation in Gyeongbuk and Ulsan.

**Conclusion:** Hearing screening tests on newborns of low income families issued with a free coupon in the southeastern area were conducted but the referred newborns after the screening tests were not well linked to the confirmation test hospitals, and hearing rehabilitation was generally not performed well after the hearing loss was confirmed. Therefore, newborns with a handicap in those areas could not develop their hearing abilities.

**Daneshi A, Mirsalehi M, Hashemi SB, Ajalloueyan M, Rajati M, Ghasemi MM, Emamdjomeh H, Asghari A, Mohammadi S, Mohseni M, Mohebbi S, Farhadi M.**

[Cochlear implantation in children with auditory neuropathy spectrum disorder: A multicenter study on auditory performance and speech production outcomes. Int J Pediatr Otorhinolaryngol.](#)

2018 May;108:12-16. doi: 10.1016/j.ijporl.2018.02.004. Epub 2018 Feb 7.

**OBJECTIVES:** To evaluate the auditory performance and speech production outcome in children with auditory neuropathy spectrum disorder (ANSD). The effect of age on the outcomes of the surgery at the time of implantation was also evaluated.

**METHODS:** Cochlear implantation was performed in 136 children with bilateral severe-to-profound hearing loss due to ANSD, at four tertiary academic centers. The patients were divided into two groups based on the age at the time of implantation; Group I: Children  $\leq 24$  months, and Group II: subjects  $>24$  months. The categories of auditory performance (CAP) and speech intelligibility rating (SIR) scores were evaluated after the first and second years of implantation. The differences between the CAP and SIR scores in the two groups were assessed.

**RESULTS:** The median CAP scores improved significantly after the cochlear implantation in all the patients ( $p$  value  $< 0.001$ ). The improvement in the CAP scores during the first year in Group II was greater than Group I ( $p$  value: 0.007), but the improvement in CAP scores tended to be significantly higher in patients who were implanted at  $\leq 24$  months ( $p$  value  $< 0.001$ ). There was no significant difference between two groups in SIR scores at first-year and second-year follow-ups. The evaluation of the SIR improvement revealed significantly higher values for Group I during the second-year follow-up ( $p$  value: 0.003).

**CONCLUSION:** The auditory performance and speech production skills of the children with ANSD improved significantly after cochlear implantation, and this improvement was affected by age at the time of implantation.

**Deshpande AK, Tan L, Lu LJ, Altaye M, Holland SK.**

[fMRI as a Preimplant Objective Tool to Predict Children's Postimplant Auditory and Language Outcomes as Measured by Parental Observations. J Am Acad Audiol.](#)

2018 May;29(5):389-404. doi: 10.3766/jaaa.16149.

**BACKGROUND:** The trends in cochlear implantation candidacy and benefit have changed rapidly in the last two decades. It is now widely accepted that early implantation leads to better postimplant outcomes. Although some generalizations can be made about postimplant auditory and language performance, neural mechanisms need to be studied to predict individual prognosis.

**PURPOSE:** The aim of this study was to use functional magnetic resonance imaging (fMRI) to identify preimplant neuroimaging biomarkers that predict children's postimplant auditory and language outcomes as measured by parental observation/reports.

**RESEARCH DESIGN:** This is a pre-post correlational measures study.

**STUDY SAMPLE:** Twelve possible cochlear implant candidates with bilateral severe to profound hearing loss were recruited via referrals for a clinical magnetic resonance imaging to ensure structural integrity of the auditory nerve for implantation.

**INTERVENTION:** Participants underwent cochlear implantation at a mean age of 19.4 mo. All children used the advanced combination encoder strategy (ACE, Cochlear Corporation™, Nucleus® Freedom cochlear implants). Three participants received an implant in the right ear; one in the left ear whereas eight participants received bilateral implants. Participants' pre-implant neuronal activation in response to two auditory stimuli was studied using an event-related fMRI method.

**DATA COLLECTION AND ANALYSIS:** Blood oxygen level dependent contrast maps were calculated for speech and noise stimuli. The general linear model was used to create z-maps. The Auditory Skills Checklist (ASC) and the SKI-HI Language Development Scale (SKI-HI LDS) were administered to the parents 2 yr after implantation. A nonparametric correlation analysis was implemented between preimplant fMRI activation and postimplant auditory and language outcomes based on ASC and SKI-HI LDS. Statistical Parametric Mapping software was used to create regression maps between fMRI activation and scores on the aforementioned tests. Regression maps were overlaid on the Imaging Research Center infant template and visualized in MRIcro.

**RESULTS:** Regression maps revealed two clusters of brain activation for the speech versus silence contrast and five clusters for the noise versus silence contrast that were significantly correlated with the parental reports. These clusters included auditory and extra-auditory regions such as the middle temporal gyrus, supramarginal gyrus, precuneus, cingulate gyrus, middle frontal gyrus, subgyral, and middle occipital gyrus. Both positive and negative correlations were observed. Correlation values for the different clusters ranged from -0.90 to 0.95 and were significant at a corrected  $p$  value of  $< 0.05$ . Correlations suggest that postimplant performance may be predicted by activation in specific brain regions.

**CONCLUSIONS:** The results of the present study suggest that (1) fMRI can be used to identify neuroimaging biomarkers of auditory and language performance before implantation and (2) activation in certain brain regions may be predictive of postimplant auditory and language performance as measured by parental observation/reports.

Dickinson LJ, Nimmo M, Morton RP, Purdy SC.

[‘Asymptomatic’ South Auckland preschool children have significant hearing loss and middle ear disease. Int J Pediatr Otorhinolaryngol.](#)

2018 Nov;114:106-110. doi: 10.1016/j.ijporl.2018.08.034. Epub 2018 Aug 30.

**BACKGROUND:** Seven hundred children were recalled for hearing screening at age 2-3 years due to a problem with their newborn hearing screen. They had all been well babies with no identified risk factors for hearing loss and hence were not scheduled for targeted follow-up to retest hearing.

**METHODS:** There were 485 children (69%) that attended the recall. The average age was 36 months (SD 3.7). Family ethnicity was Pacific Island (36%), Asian (26%), NZ European (13%), and Māori (11%), and there was a high level of deprivation in the study population. Children were screened using distortion product otoacoustic emission (DPOAE) and a parent or caregiver completed a 14-item questionnaire about ear health. The children that did not pass screening were given appointments for audiology testing. Children with hearing loss and/or middle ear problems were referred for otolaryngology review and further hearing assessments.

**RESULTS:** About one third (36%; n = 176) of children did not pass DPOAE screening; 82 (17%) had abnormal type B tympanograms and hearing loss; 29 underwent insertion of ventilation tubes, and one had a perforated tympanic membrane. There was a significant association between failed tympanometry and hearing loss (Chi-squared = 16.67, p < .001). Five children had permanent sensorineural hearing loss (SNHL), two of whom required cochlear implants for idiopathic hearing loss, with no specific risk factors. Overall 380 of 485 children screened were deemed to have normal hearing (i.e. 22% failed hearing). From the questionnaire, 15% of the caregivers with no suspicion of hearing problems did have children with significant hearing loss. Regression analysis showed that Pacific/Māori ethnicity was significantly associated with risk of hearing loss, together with questionnaire items identifying hearing problems and breathing problems.

**CONCLUSIONS:** There is a high proportion of children in South Auckland with unsuspected hearing loss; a different approach to hearing screening is warranted for this population with high rates of middle ear disease at age 3.

Elalfy M, El-Farrash R, Taha H, Ismail E, Mokhtar N.

[Auditory brainstem response in full term neonates born to mothers with iron deficiency anemia: relation to disease severity. J Matern Fetal Neonatal Med.](#)

2018 Oct 8:1-181. doi: 10.1080/14767058.2018.1533940. [Epub ahead of print]

**BACKGROUND:** Iron is crucial for fetal brain development; however, there are insufficient data regarding the effects of maternal iron deficiency anemia (IDA) on auditory neural maturation.

**AIM:** We evaluated the effect of maternal IDA on auditory brainstem response (ABR) in full term neonates.

**METHODS:** Out of 223 pregnant women, 50 were diagnosed as having IDA and 50 healthy mothers were enrolled as controls. ABR test was done for the studied neonates within 48 hours after birth and at 3 months.

**RESULTS:** We found that hemoglobin and iron profile were lower in neonates born to anemic mothers compared with controls. Of 100 neonates screened for ABR, 25 failed the test (all of them were born to anemic mothers). The majority of neonates who failed the screening ABR test (88%) had latent iron deficiency (cord blood ferritin 11-75 µg/L). After 3 months, 85 neonates underwent diagnostic ABR test which revealed significantly prolonged interpeak latencies I-III, III-V, and I-V among neonates born to IDA mothers compared with the control group. Within the IDA group, all interpeak latencies were more prolonged in neonates with latent iron deficiency and in those born to mothers with serum ferritin < 15 µg/L. Logistic regression analysis showed that maternal hemoglobin and mean corpuscular volume could predict neonatal ABR results.

**CONCLUSIONS:** IDA during late pregnancy adversely affects cord blood iron and hearing status. ABR results are closely related to the severity of maternal and neonatal iron status. Antenatal screening of pregnant mothers is needed to improve fetal iron status and prevent abnormal auditory maturation.

EI-Hattab AW, Almannai M, Sutton VR.

[Newborn Screening: History, Current Status, and Future Directions. Pediatr Clin North Am.](#)

2018 Apr;65(2):389-405. doi: 10.1016/j.pcl.2017.11.013. Epub 2017 Dec 28.

Newborn screening programs aim to achieve presymptomatic diagnosis of treatable disorders allowing for early initiation of medical care to prevent or reduce significant morbidity and mortality. Many of the conditions included in the newborn screening panels are inborn errors of metabolism; however, screening for endocrine, hematologic, immunologic, and cardiovascular diseases, and hearing loss is also included in many panels. Newborn screening tests are not diagnostic and therefore diagnostic testing is needed to confirm or exclude the suspected diagnosis. Further advancement in technology is expected to allow continuous expansion of newborn screening.

Fandiño-Cárdenas M, Idrovo AJ, Velandia R, Molina-Franky J, Alvarado-Socarras JL.

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

2018 Sep 25. doi: 10.1093/tropej/fmy055. [Epub ahead of print]

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

**Methods:** This is a cases report. The information was collected and recorded in a database between January 2016 and April 2018. We perform two auditory tests to 3 and 24 months of life. The study was developed in Aguachica (Cesar, Colombia). It is considered a high-risk area for ZIKV infection. Participants included children of mothers with confirmed ZIKV during pregnancy

or suspicious of congenital ZIKV exposure of ZIKV infection during an epidemic period in a tropical area. We defined a positive case according to the epidemiological definition and clinical criteria based on maternal symptoms. However, other children of mothers without clinical signs of Zika were evaluated at the same time. The main outcome was the presence of sensorineural hearing loss.

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

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

**Fasunla AJ.**

[Long-term effects of Maternal HIV Infection and Anti-Retroviral Medications on the Hearing of HIV-Exposed Infants. West Afr J Med.](#)

2018 May-Aug;35(2):90-96.

**BACKGROUND:** The long-term effect of maternal HIV and antiretroviral medication on the hearing of HIV-exposed infants has not been well studied. We determined and compared the hearing thresholds of HIV-exposed infants with that of HIV-unexposed infants within the first month of life and at three, six and nine months of life.

**MATERIALS AND METHODS:** This was a Case control study of 126 HIV-exposed newborns and 121 HIV-unexposed newborns. Data collected included Socio-demographic, clinical characteristics and risk factors for hearing loss. Hearing was evaluated at newborn, 3, 6, and 9 months of life. Within and between groups analyses were done with appropriate statistics. Level of significance was  $P < 0.05$ .

**RESULTS:** In both ears, the mean hearing thresholds of HIV-exposed infants were greater than those of the HIV-unexposed infants at baseline, 3, 6 and 9 months ( $P > 0.05$ ). In both groups, there was a decline in the mean hearing thresholds from baseline (new born) till 6 months of age. The highest mean threshold was recorded at 9 months. The mean hearing thresholds of infants at 3, 6, and 9 months were lower for HIV-exposed infants and higher for HIV-unexposed infants than the corresponding mean hearing thresholds measured at baseline. There was a significant strong correlation among hearing thresholds at 3, 6 and 9 months but weakly correlated with hearing thresholds at baseline.

**CONCLUSION:** There was a tendency towards higher hearing thresholds in HIV-exposed infants than the HIV-unexposed infants throughout the infancy period. This appears to have association with in-utero exposure to HIV.

**Findlen UM, Hounam GM, Alexy E, Adunka OF.**

[Early Hearing Detection and Intervention: Timely Diagnosis, Timely Management. Ear Hear.](#)

2018 Aug 21. doi: 10.1097/AUD.0000000000000647. [Epub ahead of print]

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

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

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

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

**Fitzpatrick EM, Coyle D, Gaboury I, Durieux-Smith A, Whittingham J, Grandpierre V, Na E, Salamatmanesh M.**  
[Service Preferences of Parents of Children With Mild Bilateral or Unilateral Hearing Loss: A Conjoint Analysis Study. Ear Hear.](#)

2018 Aug 20. doi: 10.1097/AUD.0000000000000651. [Epub ahead of print]

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

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



link) to identify attributes of interest for the respondents. Characteristics of the children were entered into the model to control for differences in age of diagnosis, sex, laterality of hearing loss, and hearing aid use.

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

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

**Franchella S, Bovo R, Bandolin L, Gheller F, Montino S, Borsetto D, Ghiselli S, Martini A.**

[Surgical timing for bilateral simultaneous cochlear implants: When is best? Int J Pediatr Otorhinolaryngol.](#)

2018 Jun;109:54-59. doi: 10.1016/j.ijporl.2018.03.019. Epub 2018 Mar 24.

**INTRODUCTION:** Hearing loss is considered the most common congenital disease and the prevalence of neonatal deafness can be estimated between 1 and 2 cases per 1000 live births. Infant deafness must be diagnosed as early as possible and an effective therapeutic intervention needs to be carried out in order to avoid the serious consequences of hearing deprivation during the evolutionary period: alterations in the development of central auditory pathways and lack of language acquisition. The cochlear implant (CI) has proved to be the best instrument to solve the problem of auditory deprivation. In particular, the bilateral CI gives the patient access to binaural hearing which results in benefits in terms of sound localisation and discrimination. The optimal age of application of the CI is a widely discussed topic in the scientific community and the current guidelines indicate a period between 12 and 24 months of age, even though the supporters of the application before 12 months of age are nowadays increasing.

**MATERIALS AND METHODS:** The study is observational, retrospective, monocentric. 49 paediatric patients (<18 years) with simultaneous bilateral CIs were included. The audiometric threshold and speech tests were carried out during the follow-up 3, 6 and 12 months after the CIs activation and when the patient reached 2 years of age.

**RESULTS:** The statistical analysis showed that undergoing bilateral implantation surgery before 2 years of age allows a satisfactory audiometric performance, while there are no particular benefits in performing the surgery before 1 year of age. As far as the speech outcome is concerned, the statistical analysis didn't show significant correlation between the earlier age of implantation and better speech performance if the operation is carried out before 2.5 years of age.

**CONCLUSIONS:** The results of the study indicate that the optimal age to perform the simultaneous bilateral CIs surgery is between 12 and 24 months, without demonstrating any particular benefit in carrying out the procedure before 1 year of age. This may be clinically relevant in terms of avoiding the risks of diagnostic mistakes and reducing the related surgical risk in children under 1 year of age.

**Gardner PA, Shanley R, Perry BP.**

[Failure rate in pediatric cochlear implantation and hearing results following revision surgery. Int J Pediatr Otorhinolaryngol.](#)

2018 Aug;111:13-15. doi: 10.1016/j.ijporl.2018.05.017. Epub 2018 May 24.

**INTRODUCTION:** In a discussion of the risks and benefits of pediatric cochlear implantation, device failure and the need for revision surgery is often overlooked. The failure rate has not been investigated extensively for this population of patients. Hearing results are under-reported following revision surgery as well. We will review our experience with cochlear implant failure, revision, and hearing results when available to better guide the preoperative counseling of families considering cochlear implantation.

**METHODS:** Retrospective chart review of all children undergoing cochlear implantation from 2004 to 2014.

**RESULTS:** In this review of 579 cases of pediatric cochlear implantation, a 4.7% device failure rate was identified. Additionally, there was a 0.3% device infection rate, as well as a 0.3% electrode extrusion rate. 10 patients had audiometric data prior to and following revision surgery. These data demonstrate similar pre-failure and post revision surgery pure tone average.

**CONCLUSION:** Cochlear implant device failure is the most common long-term complication of surgery; fortunately, hearing outcomes following revision surgery, evaluated with pure tone average, revealed no decline in auditory performance.

**Garg S, Kohli C, Mangla V, Chadha S, Singh MM, Dahiya N.**

[An Epidemiological Study on Burden of Hearing Loss and Its Associated Factors in Delhi, India. Ann Otol Rhinol Laryngol.](#)

2018 Sep;127(9):614-619. doi: 10.1177/0003489418781968. Epub 2018 Jun 25.

**INTRODUCTION:** Hearing loss is the second most common cause of years lived with disability (YLD). The present study was conducted with an objective to determine the prevalence, severity, and sociodemographic correlates of hearing loss among people aged 3 months and above in selected areas of Delhi, India.

**MATERIAL AND METHODS:** A community-based cross-sectional study was conducted in selected rural and urban areas of Delhi among population aged 3 months and above. Of the total sample size of 664, 85 study subjects (17 households) were taken from the rural area, and 579 (116 households) were taken from urban areas. The hearing test and ear examination was carried out using handheld oto-acoustic emission (OAE) in children <5 years of age and pure tone audiometry in individuals

above 5 years of age. SPSS software was used for data analysis. Chi-square test was used to analyze difference between proportions.

**RESULTS:** Overall prevalence of hearing loss was 25.1%. Conductive hearing loss was present among 61 (10.3%) subjects, mixed hearing loss was found among 5 (0.8%) subjects, and sensorineural hearing loss among 94 (15.8%) subjects. On OAE, 62 (89.9%) children passed the test, and 7 (10.1%) were referred. Increasing age, female gender, and low education were significantly associated with hearing loss.

**CONCLUSION:** There was high prevalence of hearing loss in the study sample. Urgent interventions are required to identify individuals with hearing loss so that its serious complications can be reduced.

**Garinis AC, Kempf A, Tharpe AM, Weitkamp JH, McEvoy C, Steyger PS.**

[Monitoring neonates for ototoxicity. \*Int J Audiol.\*](#)

2018 Sep;57(sup4):S41-S48. doi: 10.1080/14992027.2017.1339130. Epub 2017 Jun 22.

**OBJECTIVES:** Neonates admitted to the neonatal intensive care unit (NICU) are at greater risk of permanent hearing loss compared to infants in well mother and baby units. Several factors have been associated with this increased prevalence of hearing loss, including congenital infections (e.g. cytomegalovirus or syphilis), ototoxic drugs (such as aminoglycoside or glycopeptide antibiotics), low birth weight, hypoxia and length of stay. The aetiology of this increased prevalence of hearing loss remains poorly understood.

**DESIGN:** Here we review current practice and discuss the feasibility of designing improved ototoxicity screening and monitoring protocols to better identify acquired, drug-induced hearing loss in NICU neonates.

**STUDY SAMPLE:** A review of published literature.

**CONCLUSIONS:** We conclude that current audiological screening or monitoring protocols for neonates are not designed to adequately detect early onset of ototoxicity. This paper offers a detailed review of evidence-based research, and offers recommendations for developing and implementing an ototoxicity monitoring protocol for young infants, before and after discharge from the hospital.

**George L, Patel JB, Park N, Manimtim WM.**

[Implementation of Updated Hearing Screen Guidelines in a Level IV NICU-A Quality Improvement Project. \*Am J Perinatol.\*](#)

2018 Aug 6. doi: 10.1055/s-0038-1668170. [Epub ahead of print]

Hearing loss is the most common congenital birth defect. In 2007, American Academy of Pediatrics updated the hearing screen guidelines to recommend hearing screen by 1 month of age, diagnostic evaluation by 3 months, and early interventions by 6 months. Early interventions have been shown to improve developmental outcome in children with hearing loss. Infants admitted to the neonatal intensive care unit (NICU) are at higher risk for hearing loss. For infants born before 34 weeks' gestation, there are no guidelines for initial hearing screen. Although auditory brain stem response can be reliably performed at 32 to 34 weeks, in most NICUs, they are screened prior to discharge per universal hearing screen guidelines. In high-risk infants, often with prolonged hospitalization, this leads to missed opportunity for early detection and implementation of early intervention services. Using quality improvement methodology, an updated hearing screen algorithm was developed and implemented in our level IV NICU along with an electronic medical record tool to improve the process of identifying infants meeting criteria for hearing screen.

**Goh LC, Azman A, Siti HBK, Khoo WV, Muthukumarasamy PA, Thong MK, Abu Bakar Z, Manuel AM.**

[An audiological evaluation of syndromic and non-syndromic craniosynostosis in pre-school going children. \*Int J Pediatr Otorhinolaryngol.\*](#)

2018 Jun;109:50-53. doi: 10.1016/j.ijporl.2018.03.010. Epub 2018 Mar 14.

**OBJECTIVE:** To study the audiological outcome and early screening of pre-school going children with craniosynostosis under follow-up at the University of Malaya Medical Center (UMMC), Kuala Lumpur, Malaysia over a 10 year period.

**METHODS:** A retrospective descriptive cohort study on the audiological findings detected during the first hearing assessment done on a child with craniosynostosis using otoacoustic emissions, pure tone audiometry or auditory brainstem response examination. The main aim of this study was to evaluate the type and severity of hearing loss when compared between syndromic and non-syndromic craniosynostosis, and other associated contributory factors.

**RESULTS:** A total of 31 patients with 62 ears consisting of 14 male patients and 17 female patients were evaluated. Twenty two patients (71%) were syndromic and 9 (29%) were non-syndromic craniosynostosis. Amongst the syndromic craniosynostosis, 9 (41%) had Apert syndrome, 7 (32%) had Crouzon syndrome, 5 (23%) had Pfeiffer syndrome and 1 (4%) had Shaethre Chotzen syndrome. Patients with syndromic craniosynostosis were more likely to present with all types and severity of hearing loss, including severe to profound sensorineural hearing loss while children with non-syndromic craniosynostosis were likely to present with normal hearing ( $p < 0.05$ ). In addition, when the first hearing test was done at a later age, a hearing loss including sensorineural hearing loss is more likely to be present in a child with syndromic craniosynostosis ( $p < 0.05$ ).

**CONCLUSION:** Our study suggested that children who are born with syndromic craniosynostosis were more likely to suffer from a hearing loss, including that of a severe to profound degree compared to children with non-syndromic craniosynostosis. In addition to that, hearing loss is more likely to be detected when the first hearing test is done at a later age, and this can be an irreversible sensorineural hearing loss. We would like to advocate the need for early audiological screening and follow up in children with syndromic craniosynostosis.

Grosse SD, Mason CA, Gaffney M, Thomson V, White KR.

[What Contribution Did Economic Evidence Make to the Adoption of Universal Newborn Hearing Screening Policies in the United States? \*Int J Neonatal Screen\*.](#)

2018;4(3):25. doi: 10.3390/ijns4030025. Epub 2018 Jul 20.

Universal newborn hearing screening (UNHS), when accompanied by timely access to intervention services, can improve language outcomes for children born deaf or hard of hearing (D/HH) and result in economic benefits to society. Early Hearing Detection and Intervention (EHDI) programs promote UNHS and using information systems support access to follow-up diagnostic and early intervention services so that infants can be screened no later than 1 month of age, with those who do not pass their screen receiving diagnostic evaluation no later than 3 months of age, and those with diagnosed hearing loss receiving intervention services no later than 6 months of age. In this paper, we first document the rapid roll-out of UNHS/EHDI policies and programs at the national and state/territorial levels in the United States between 1997 and 2005. We then review cost analyses and economic arguments that were made in advancing those policies in the United States. Finally, we examine evidence on language and educational outcomes that pertain to the economic benefits of UNHS/EHDI. In conclusion, although formal cost-effectiveness analyses do not appear to have played a decisive role, informal economic assessments of costs and benefits appear to have contributed to the adoption of UNHS policies in the United States.

Hansashree YS, Bhatt SH, Nimbalkar S, Mishra G.

[Non-compliance With Neonatal Hearing Screening Follow-up in Rural Western India. \*Indian Pediatr\*.](#)

2018 Jun 15;55(6):482-484.

**OBJECTIVE:** The reasons of failure to follow-up for the Universal Neonatal Hearing Screening (UNHS) program were delineated.

**METHODS:** Review of case records for data related to follow-up of neonates who underwent the UNHS between February 2012 - January 2015.

**RESULTS:** 2534 neonates underwent primary screening with Distortion Product Oto-acoustic Emission (DPOAE). 14 (26.9%) were lost to follow-up between the first and second DPOAE screenings. 275 neonates (including high-risk cases) were to undergo confirmatory Brain Evoked Response Audiometry testing out of which 201 (73.4%) came for follow-up. Out of 74 who failed to follow-up (including those lost between first and second DOPAE screenings), unwillingness and non-compliance was the commonest reason.

**CONCLUSION:** Increasing awareness and counseling of the caretaker are important interventions for ensuring good follow-up in hearing screening programs.

Hao Z, Fu D, Ming Y, Yang J, Huang Q, Lin W, Zhang H, Zhang B, Zhou A, Hu X, Yao C, Dong Y, Ring HZ, Ring BZ. [Large scale newborn deafness genetic screening of 142,417 neonates in Wuhan, China. \*PLoS One\*.](#)

2018 Apr 10;13(4):e0195740. doi: 10.1371/journal.pone.0195740. eCollection 2018.

Almost one third of the three million people in China suffering severe deafness are children, and 50% of these cases are believed to have genetic components to their etiology. Newborn hearing genetic screening can complement Universal Neonatal Hearing Screening for the diagnosis of congenital hearing loss as well as identifying children at risk for late-onset and progressive hearing impairment. The aim of this joint academic and Ministry of Health project was to prototype a cost effective newborn genetic screen in a community health setting on a city-wide level, and to ascertain the prevalence of variation at loci that have been associated with non-syndromic hearing loss. With the participation of 143 local hospitals in the city of Wuhan, China we screened 142,417 neonates born between May 2014 and Dec. 2015. The variants GJB2 c.235delC, SLC26A4 c.919-2A>G, and mitochondrial variants m.1555A>G and m.1494C>T were assayed using real time PCR. Newborns found to carry a variant were re-assayed by sequencing in duplicate. Within a subset of 707 newborns we assayed using real-time PCR and ARMS-PCR to compare cost, sensitivity and operating procedure. The most frequent hearing loss associated allele detected in this population was the 235delC variant in GJB2 gene. In total, 4289 (3.01%) newborns were found to carry at least one allele of either GJB2 c.235delC, SLC26A4 c.919-2A>G or two assayed MT-RNR1 variants. There was complete accordance between the real-time PCR and the ARMS PCR, though the real-time PCR had a much lower failure rate. Real-time PCR had a lower cost and operating time than ARMS PCR. Ongoing collaboration with the participating hospitals will determine the specificity and sensitivity of the association of the variants with hearing loss at birth and arising in early childhood, allowing an estimation of the benefits of newborn hearing genetic screening in a large-scale community setting.

Heffernan CB, McKeon MG, Molony S, Kawai K, Stiles DJ, Lachenauer CS, Kenna MA, Watters K.

[Does Clarithromycin Cause Hearing Loss? A 12-Year Review of Clarithromycin Therapy for Nontuberculous Mycobacterial Lymphadenitis in Children. \*Ann Otol Rhinol Laryngol\*.](#)

2018 Oct;127(10):687-693. doi: 10.1177/0003489418788112. Epub 2018 Jul 21.

**OBJECTIVE(S):** The objective was to describe the characteristics of hearing losses documented in patients treated with clarithromycin alone for nontuberculous mycobacterial NTM lymphadenitis in a pediatric tertiary care center over a 12-year period.

**METHODS:** An institutional review board (IRB) approval was obtained. A database search was performed using the ICD-10 diagnosis codes 31.0, 31.1, and 31.8 between January 2004 and January 2017. A REDCap database was created to record variables. Patients were included if they received clarithromycin alone and had, at the minimum, a baseline audiology assessment, and 1 further evaluation during treatment. Fisher's exact test was used to analyze categorical variables, and Wilcoxon rank sum test was used to analyze continuous variables.

**RESULTS:** A total of 167 patients with cervicofacial NTM were identified. Of them, 42 patients fulfilled inclusion criteria. Three children (7%) developed a hearing loss (HL) between 25 and 63 days after starting treatment. HL was unilateral in 2 children. HL persisted in 1 child following cessation of treatment. However, this patient had Rubinstein Taybi syndrome, limiting our ability to attribute the HL solely to clarithromycin.

**CONCLUSION:** We noted a 7% hearing loss rate in our series. Confounding issues, such as 1 patient with a syndrome potentially contributing to HL, and limitations to this study, including retrospective design and loss to follow-up, temper our ability to conclude that clarithromycin was the sole cause of these HL. However, enough supporting data for a role in clarithromycin causing HL exist that testing should be considered for patients undergoing long-term clarithromycin treatment.

**Hilditch C1, Liersch B, Spurrier N, Callander EJ, Cooper C, Keir AK.**

[Does screening for congenital cytomegalovirus at birth improve longer term hearing outcomes? Arch Dis Child.](#)

2018 Oct;103(10):988-992. doi: 10.1136/archdischild-2017-314404. Epub 2018 Apr 28.

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

**Huang B, Han M, Wang G, Huang S, Zeng J, Yuan Y, Dai P.**

[Genetic mutations in non-syndromic deafness patients in Hainan Province have a different mutational spectrum compared to patients from Mainland China. Int J Pediatr Otorhinolaryngol.](#)

2018 May;108:49-54. doi: 10.1016/j.ijporl.2018.02.015. Epub 2018 Feb 15.

**OBJECTIVES:** To provide appropriate genetic testing and counseling for non-syndromic hearing impairment patients in Hainan Province, an island in the South China Sea.

**METHODS:** 299 unrelated students with non-syndromic hearing loss who attended a special education school in Hainan Province were enrolled in this study. Three prominent deafness-related genes (GJB2, SLC26A4, and mtDNA 12S rRNA) were analyzed using Sanger sequencing.

**RESULTS:** GJB2 mutations were detected in 32.78% (98/299) of the entire cohort; however, only 5.69% (17/299) had two confirmed pathogenic mutations. The most common mutation observed in this population was c.109G > A in the GJB2 gene, with an allelic frequency of 15.05% (90/598), which is significantly higher than that reported in previous cohorts. A total of 16 patients had two confirmed pathogenic SLC26A4 gene mutations, and 16 patients had one. The IVS7-2A > G mutation was the most commonly observed, with an allelic frequency of 3.51% (21/598). Three patients had a m.1555A > G mutation in the mtDNA 12S rRNA gene.

**CONCLUSIONS:** These results reveal that genetic etiology occurred in 11.71% (35/299) of patients, suggesting that Hainan province have a different mutational spectrum compare to Mainland China in non-syndromic deafness patients, which provide useful information to genetic counseling in Hainan province.

**Huang Z, Gordish-Dressman H, Preciado D, Reilly BK.**

[Pediatric cochlear implantation: Variation in income, race, payer, and charges across five states. Laryngoscope.](#)

2018 Apr;128(4):954-958. doi: 10.1002/lary.26686. Epub 2017 Jun 9.

**OBJECTIVES/HYPOTHESIS:** Our objectives were to investigate pediatric cochlear implantation (PCI) across representative states within the United States and analyze any geographical differences in age, median household income, race, insurance, and total medical charges.

**STUDY DESIGN:** Cross-sectional.

**METHODS:** Data from children (aged 0.5-18 years) who received cochlear implantation surgery were collected from the 2011 State Ambulatory Surgery and Services Databases from California (CA), Florida (FL), Maryland (MD), New York (NY), and Kentucky (KY) as a part of the Healthcare Cost and Utilization Project. We performed data analysis using a combination of Kruskal-Wallis and Wilcoxon rank sum tests, as well as nominal logistic regression.

**RESULTS:** Five hundred twelve cases of PCI were performed during 2011 across the five states. The overall mean and median age of implantation were 5.6 years and 4 years, respectively. There was no statistical difference in age of implantation across states ( $P = .85$ ). However, there were statistical differences in primary payer ( $P < .001$ ), median household income quartiles of patients who received an implant ( $P < .006$ ), race ( $P < .001$ ), and total median hospital charges for four of the states, with the exception of CA ( $P < .001$ ).

**CONCLUSIONS:** Age of PCI appears to be similar across the five states in cross-sectional analysis. Geographic variations in charges, payer, race, and median household income occur with statistical significance in PCI. Further analysis of contributing factors at each state level may help elucidate the root cause of these disparities and improve and justify a uniform approach to healthcare delivery and standards of care.

**Hunter LL, Blankenship CM, Gunter RG, Keefe DH, Feeney MP, Brown DK, Baroch K.**

[Cochlear Microphonic and Summating Potential Responses from Click-Evoked Auditory Brain Stem Responses in High-Risk and Normal Infants. J Am Acad Audiol.](#)

2018 May;29(5):427-442. doi: 10.3766/jaaa.17085.

**BACKGROUND:** Examination of cochlear and neural potentials is necessary to assess sensory and neural status in infants, especially those cared for in neonatal intensive care units (NICU) who have high rates of hyperbilirubinemia and thus are at



risk for auditory neuropathy (AN).

**PURPOSE:** The purpose of this study was to determine whether recording parameters commonly used in click-evoked auditory brain stem response (ABR) are useful for recording cochlear microphonic (CM) and Wave I in infants at risk for AN. Specifically, we analyzed CM, summing potential (SP), and Waves I, III, and V. The overall aim was to compare latencies and amplitudes of evoked responses in infants cared for in NICUs with infants in a well-baby nursery (WBN), both of which passed newborn hearing screening.

**RESEARCH DESIGN:** This is a prospective study in which infants who passed ABR newborn hearing screening were grouped based on their birth history (WBN and NICU). All infants had normal hearing status when tested with diagnostic ABR at about one month of age, corrected for prematurity.

**STUDY SAMPLE:** Thirty infants (53 ears) from the WBN [mean corrected age at test = 5.0 weeks (wks.)] and thirty-two infants (59 ears) from the NICU (mean corrected age at test = 5.7 wks.) with normal hearing were included in this study. In addition, two infants were included as comparative case studies, one that was diagnosed with AN and another case that was diagnosed with bilateral sensorineural hearing loss (SNHL).

**DATA COLLECTION AND ANALYSIS:** Diagnostic ABR, including click and tone-burst air- and bone-conduction stimuli were recorded. Peak Waves I, III, and V; SP; and CM latency and amplitude (peak to trough) were measured to determine if there were differences in ABR and electrocochleography (ECoChG) variables between WBN and NICU infants.

**RESULTS:** No significant group differences were found between WBN and NICU groups for ABR waveforms, CM, or SP, including amplitude and latency values. The majority (75%) of the NICU group had hyperbilirubinemia, but overall, they did not show evidence of effects in their ECoChG or ABR responses when tested at about one-month corrected age. These data may serve as a normative sample for NICU and well infant ECoChG and ABR latencies at one-month corrected age. Two infant case studies, one diagnosed with AN and another with SNHL demonstrated the complexity of using ECoChG and otoacoustic emissions to assess the risk of AN in individual cases.

**CONCLUSIONS:** CM and SPs can be readily measured using standard click stimuli in both well and NICU infants. Normative ranges for latency and amplitude are useful for interpreting ECoChG and ABR components. Inclusion of ECoChG and ABR tests in a test battery that also includes otoacoustic emission and acoustic reflex tests may provide a more refined assessment of the risks of AN and SNHL in infants.

**Ismail AI, Abdul Majid AH, Zakaria MN, Abdullah NAC, Hamzah S4, Mukari SZS.**

**Factors predicting health practitioners' awareness of UNHS program in Malaysian non-public hospitals. *Int J Pediatr Otorhinolaryngol.***

2018 Jun;109:78-84. doi: 10.1016/j.ijporl.2018.03.030. Epub 2018 Mar 28.

**OBJECTIVE:** The current study aims to examine the effects of human resource (measured with the perception of health workers' perception towards UNHS), screening equipment, program layout and screening techniques on healthcare practitioners' awareness (measured with knowledge) of universal newborn hearing screening (UNHS) in Malaysian non-public hospitals.

**METHODS:** Via cross sectional approach, the current study collected data using a validated questionnaire to obtain information on the awareness of UNHS program among the health practitioners and to test the formulated hypotheses. 51, representing 81% response rate, out of 63 questionnaires distributed to the health professionals were returned and usable for statistical analysis. The survey instruments involving healthcare practitioners' awareness, human resource, program layout, screening instrument, and screening techniques instruments were adapted and scaled with 7-point Likert scale ranging from 1 (little) to 7 (many). Partial Least Squares (PLS) algorithm and bootstrapping techniques were employed to test the hypotheses of the study.

**RESULTS:** With the result involving beta values, t-values and p-values (i.e.  $\beta=0.478$ ,  $t=1.904$ ,  $p<0.10$ ;  $\beta=0.809$ ,  $t=3.921$ ,  $p<0.01$ ;  $\beta=-0.436$ ,  $t=1.870$ ,  $p<0.10$ ), human resource, measured with training, functional equipment and program layout, are held to be significant predictors of enhanced knowledge of health practitioners. Likewise, program layout, human resource, screening technique and screening instrument explain 71% variance in health practitioners' awareness. Health practitioners' awareness is explained by program layout, human resource, and screening instrument with effect size ( $f^2$ ) of 0.065, 0.621, and 0.211 respectively, indicating that program layout, human resource, and screening instrument have small, large and medium effect size on health practitioners' awareness respectively. However, screening technique has zero effect on health practitioners' awareness, indicating the reason why T-statistics is not significant.

**CONCLUSION:** Having started the UNHS program in 2003, non-public hospitals have more experienced and well-trained employees dealing with the screening tools and instrument, and the program layout is well structured in the hospitals. Yet, the issue of homogeneity exists. Non-public hospitals charge for the service they render, and, in turn, they would ensure quality service, given that they are profit-driven and/or profit-making establishments, and that they would have no option other than provision of value-added and innovative services. The employees in the non-public hospitals have less screening to carry out, given the low number of babies delivered in the private hospitals. In addition, non-significant relationship between screening techniques and healthcare practitioners' awareness of UNHS program is connected with the fact that the techniques that are practiced among public and non-public hospital are similar and standardized. Limitations and suggestions were discussed.

Jacob-Corteletti LCB, Araújo ES, Duarte JL, Zucki F, Alvarenga KF.

[Acoustic Reflex Testing in Neonatal Hearing Screening and Subsequent Audiological Evaluation. J Speech Lang Hear Res.](#)

2018 Jul 13;61(7):1784-1793. doi: 10.1044/2018\_JSLHR-H-16-0291.

**Purpose:** The aims of the study were to examine the acoustic reflex screening and threshold in healthy neonates and those at risk of hearing loss and to determine the effect of birth weight and gestational age on acoustic stapedial reflex (ASR).

**Method:** We assessed 18 healthy neonates (Group I) and 16 with at least 1 risk factor for hearing loss (Group II); all of them passed the transient evoked otoacoustic emission test that assessed neonatal hearing. The test battery included an acoustic reflex screening with activators of 0.5, 1, 2, and 4 kHz and broadband noise and an acoustic reflex threshold test with all of them, except for the broadband noise activator.

**Results:** In the evaluated neonates, the main risk factors were the gestational age at birth and a low birth weight; hence, these were further analyzed. The lower the gestational age at birth and birth weight, the less likely that an acoustic reflex would be elicited by pure-tone activators. This effect was significant at the frequencies of 0.5, 1, and 2 kHz for gestational age at birth and at the frequencies of 1 and 2 kHz for birth weight. When the broadband noise stimulus was used, a response was elicited in all neonates in both groups. When the pure-tone stimulus was used, the Group II showed the highest acoustic reflex thresholds and the highest percentage of cases with an absent ASR. The ASR threshold varied from 50 to 100 dB HL in both groups. Group II presented higher mean ASR thresholds than Group I, this difference being significant at frequencies of 1, 2, and 4 kHz.

**Conclusions:** Birth weight and gestational age at birth were related to the elicitation of the acoustic reflex. Neonates with these risk factors for hearing impairment were less likely to exhibit the acoustic reflex and had higher thresholds.

Jaimes C, Delgado J, Cunnane MB, Hedrick HL, Adzick NS, Gee MS, Victoria T.

[Does 3-T fetal MRI induce adverse acoustic effects in the neonate? A preliminary study comparing postnatal auditory test performance of fetuses scanned at 1.5 and 3 T. \*Pediatr Radiol.\*](#)

2018 Oct 8. doi: 10.1007/s00247-018-4261-2. [Epub ahead of print]

**BACKGROUND:** Fetal MRI at 3 T is associated with increased acoustic noise relative to 1.5 T.

**OBJECTIVE:** The goal of this study is to determine if there is an increased prevalence of congenital hearing loss in neonates who had a 3-T prenatal MR vs. those who had it at 1.5 T.

**MATERIALS AND METHODS:** We retrospectively identified all subjects who had 3-T fetal MRI between 2012 and 2016 and also underwent universal neonatal hearing screening within 60 days of birth. Fetuses with incomplete hearing screening, magnetic resonance imaging (MRI) studies at both field strengths or fetuses affected by conditions associated with hearing loss were excluded. A random group of controls scanned at 1.5 T was identified. Five subjects had repeat same-strength MRIs (one at 3 T and four at 1.5 T). The pass/fail rate of the transient otoacoustic emissions test and auditory brainstem response test were compared using the Fisher exact test. A logistic regression was performed to assess the effects of other known risk factors for congenital hearing loss.

**RESULTS:** Three hundred forty fetal MRI examinations were performed at 3 T, of which 62 met inclusion criteria. A control population of 1.5-T fetal MRI patients was created using the same exclusion criteria, with 62 patients randomly selected from the eligible population. The fail rates of transient otoacoustic emissions test for the 1.5-T and 3-T groups were 9.7% and 6.5%, respectively, and for the auditory brainstem response test were 3.2% and 1.6%, respectively. There was no significant difference in the fail rate of either test between groups ( $P=0.74$  for transient otoacoustic emissions test, and  $P=0.8$  for auditory brainstem response test). The median gestational age of the 3-T group was 30 weeks, 1 day, significantly higher ( $P<0.001$ ) than the 1.5-T group (median gestational age: 20 weeks, 2 days).

**CONCLUSION:** Our findings suggest that the increase in noise associated with 3 T does not increase the rate of clinically detectable hearing abnormalities.

Jeong SW, Chung SH, Kim LS.

[P1 cortical auditory evoked potential in children with unilateral or bilateral cochlear implants; implication for the timing of second cochlear implantation. \*Eur Arch Otorhinolaryngol.\*](#)

2018 Jul;275(7):1759-1765. doi: 10.1007/s00405-018-5021-5. Epub 2018 May 31.

**OBJECTIVE:** To examine maturation of the central auditory pathway, using P1 cortical auditory evoked potential (CAEP), in children who had received unilateral or bilateral cochlear implantation (CI).

**STUDY DESIGN:** Prospective study.

**SETTING:** Tertiary referral hospital.

**METHODS:** Twenty children who had received CI due to congenital, or prelingual, deafness participated in the study. Participants had received the 1st implant at a mean age of  $3.4 \pm 0.7$  years; 16 had also received a 2nd CI for the contralateral ear, at a mean age of  $11.1 \pm 2.1$  years. P1 CAEP was recorded while using the 1st implant and, for those who received contralateral CI, within 2 weeks of switching on the 2nd implant. Relations between P1 latency and duration with the 1st implant, and between age at 1st CI and P1 latency, were investigated. Relations between P1 latency with the 1st and 2nd implants, and between the interstage interval and difference between P1 latencies with the 1st and 2nd implants, were also examined.

**RESULTS:** P1 CAEP with the 1st implant was present in 16 of the 20 children. Mean P1 latency was shorter in the early CI group compared with the late CI group, but this difference was not statistically significant ( $p=0.154$ ). There was a significant negative correlation between the duration with the 1st implant and P1 latency ( $r=-0.783$ ,  $p<0.001$ ). Among the 16 children with sequential bilateral CI, P1 CAEP with the 2nd implant was present in 10. There was a significant negative correlation

between the duration with the 1st implant before receiving the 2nd implant and P1 latency with the 2nd implant ( $r=-0.710$ ,  $p=0.021$ ); there was also a significant positive correlation between P1 latency with the 1st and 2nd implants ( $r=0.722$ ,  $p=0.018$ ). There was not a significant correlation between interstage interval and the difference between the two P1 latencies ( $r=-0.430$ ,  $p=0.248$ ).

**CONCLUSION:** Longer cochlear implant use is associated with shorter P1 latency. Unilateral hearing with the 1st implant may positively affect P1 latency with the 2nd CI ear. These findings imply that increased auditory experience may influence central auditory pathway maturation and that the degree of central auditory pathway maturation before the 2nd CI, rather than the timing when the surgery is received, may influence 2nd CI outcome in children with sequential bilateral cochlear implants.

**Jones AL, Lambert AW, Barnett M.**

[Nursing students: Training and maintaining universal newborn hearing screening knowledge. Nurse Educ Pract.](#) 2018 Sep;32:72-77. doi: 10.1016/j.nepr.2018.07.011. Epub 2018 Jul 20.

The purpose of the study was to obtain information concerning the effectiveness of a training program to equip nursing students in administering universal newborn hearing screening procedures, correctly recording results and clearly explaining outcomes to parent/guardians. First semester and third semester nursing students completed the National Center for Hearing Assessment and Management (NCHAM) Utah State University™ Universal Newborn Hearing Screening (UNHS) training as an in-class assignment. Students were surveyed and knowledge assessed prior to and post training. Overall results showed that all student benefited from the training.

**Kanji A1, Khoza-Shangase K1.**

[Objective Hearing Screening Measures: An Exploration of a Suitable Combination for Risk-Based Newborn Hearing Screening. J Am Acad Audiol.](#)

2018 Jun;29(6):495-502. doi: 10.3766/jaaa.16155.

**BACKGROUND:** The ideal hearing screening measure is yet to be defined, with various newborn hearing screening protocols currently being recommended for different contexts. Such diverse recommendations call for further exploration and definition of feasible and context-specific protocols.

**PURPOSE:** The aim of the study was to establish which combinations of audiological screening measures provide both true-positive (TP) and true-negative (TN) results for risk-based hearing screening, at and across time.

**RESEARCH DESIGN:** A longitudinal, repeated-measures design was employed.

**STUDY SAMPLE:** Three-hundred and twenty-five participants comprised the initial study sample. These participants comprised newborns and infants who were discharged from the neonatal intensive care unit and high care wards to “step down” wards at two public sector hospitals within an academic hospital complex.

**DATA COLLECTION AND ANALYSIS:** Transient evoked otoacoustic emissions (TEOAEs), distortion product otoacoustic emissions (DPOAEs), and automated auditory brainstem response (AABR) were conducted at the initial and repeat hearing screening. Diagnostic audiological assessments were also conducted. Results from combinations of audiological screening measures at the initial and repeat hearing screening were analyzed in relation to the final diagnostic outcome ( $n = 91$ ). Participants were classified as presenting with an overall “refer” if the outcome for any one test was “refer.” The overall screening outcomes for different test combinations were compared using McNemar’s test for paired data. Proportions across different test combinations were compared by the z-test for proportions.

**RESULTS:** Because of the absence of participants with hearing loss in the current study sample, analysis could only be conducted in relation to TN findings (specificity) and not TP findings (sensitivity). The percentage of TN findings was highest at the repeat hearing screening using any test or combination of tests when compared with findings from the initial hearing screening. TEOAE combined with AABR (TEOAE/AABR) ( $p < 0.0001$ ), DPOAE combined with AABR (DPOAE/AABR) ( $p < 0.0001$ ), and the combination of all three screening measures ( $p < 0.0001$ ) yielded the highest percentage specificity at the repeat hearing screening when compared with the initial hearing screening.

**CONCLUSIONS:** The best specificity was noted at the repeat hearing screening. Within a resource stricken context, where availability of all screening measures options may not be feasible, current study findings suggest the use of a two-stage AABR protocol or TEOAE/AABR protocol.

**Kanji A.**

[Early hearing detection and intervention: Reflections from the South African context. S Afr J Commun Disord.](#)

2018 Apr 19;65(1):e1-e3. doi: 10.4102/sajcd.v65i1.581.

For researchers and clinicians in developing contexts like South Africa, the establishment of universal newborn hearing screening (UNHS) programmes is something which we have strived to achieve. However, we need to ask the question as to whether we have attempted to view our ultimate goal of achieving mandated UNHS programmes from the perspective of the South African healthcare system as a whole. The current manuscript is aimed at providing an overview of audiological services within a broader context, with reflections from a South African perspective, and a suggestion to consider alternatives to UNHS, particularly in the South African public health care sector.

**Kanona H, Stephenson K, D’Arco F, Rajput K, Cochrane L, Jephson C.**

[Computed tomography versus magnetic resonance imaging in paediatric cochlear implant assessment: a pilot study and our experience at Great Ormond Street Hospital. J Laryngol Otol.](#)

2018 Jun;132(6):529-533. doi: 10.1017/S0022215118000440.

**BACKGROUND:** To date, there is a lack of consensus regarding the use of both computed tomography and magnetic reso-

nance imaging in the pre-operative assessment of cochlear implant candidates.

**METHODS:** Twenty-five patients underwent high-resolution computed tomography and magnetic resonance imaging. 'Control scores' describing the expected visualisation of specific features by computed tomography and magnetic resonance imaging were established. An independent radiological review of all computed tomography and magnetic resonance imaging scan features was then compared to the control scores and the findings recorded.

**RESULTS:** Agreement with control scores occurred in 83 per cent (20 out of 24) of computed tomography scans and 91 per cent (21 out of 23) of magnetic resonance imaging scans. Radiological abnormalities were demonstrated in 16 per cent of brain scans and 18 per cent of temporal bone investigations.

**Kaspar A, Newton O, Kei J, Driscoll C, Swanepoel W, Goulios H.**

**[Prevalence of otitis media and risk-factors for sensorineural hearing loss among infants attending Child Welfare Clinics in the Solomon Islands. Int J Pediatr Otorhinolaryngol.](#)**

2018 Aug;111:21-25. doi: 10.1016/j.ijporl.2018.05.021. Epub 2018 May 19.

**OBJECTIVES:** The present study aimed to assess the prevalence of otitis media and risk-factors for sensorineural hearing loss among infants in the Solomon Islands, in order to recommend an Infant Ear and Hearing Program that would be suitable to the Pacific Island context.

**METHODS:** Ear examinations and the JCIH Risk-Factor Questionnaire were administered to 288 infants attending Child Welfare Clinics in the Solomon Islands.

**RESULTS:** Overall, 150 infants (52.084%) presented with bilateral normal ear examinations and no risk-factors for SNHL. There were 73 infants (25.34%) with ear pathology in at least one ear, 13 (4.5%) of whom required referral to the ENT Clinic for medical management. The most common pathology was otitis media with effusion (OME) (21.87%). Infants aged 7-12 months were significantly more likely to present with OME ( $p < 0.001$ ) and a history of otitis media ( $p = 0.017$ ) than infants aged 0-6 months. There were 71 infants (24.65%) with at least one risk-factor for sensorineural hearing loss. The most common risk-factors were ototoxicity (8.3%), non-elective caesarean delivery (6.59%), and possible in-utero syphilis infection (5.55%).

**CONCLUSIONS:** The prevalence of otitis media and risk-factors for sensorineural hearing loss indicate the importance of initiating Infant Ear and Hearing Programs in the Solomon Islands. Program should facilitate early education on prevention of ear disease, as well as early diagnosis and management of children with hearing loss.

**CONCLUSION:** Assessment in the paediatric setting constitutes a special situation given the likelihood of congenital temporal bone abnormalities and associated co-morbidities that may be relevant to surgery and prognosis following cochlear implantation. Both computed tomography and magnetic resonance imaging contribute valuable information and remain necessary in paediatric cochlear implant pre-operative assessment.

**Keihanidost Z, Tabrizi A, Amini E, Sedaghat M, Ghahremani A, Shariat M, Kavyani Z.**

**[Risk Factors for Hearing Loss and Its Prevalence in Neonates Older than 6 Months with History of Hospitalization in Intensive Care Unit. Iran J Child Neurol.](#)**

2018 Fall;12(4):153-161.

**Objectives:** Hearing loss is one of the most important disabilities in neonates. Delay in the detection of hearing loss leads to impaired development and may prevent the acquisition of speech. We aimed to determine the risk factors associated with hearing loss in neonatal patients aged more than 6 months with a history of hospitalization in Neonatal Intensive Care Unit (NICU).

**Methods:** In this case-control study, screening for hearing loss was carried out on 325 neonates aged 6-12 months referred to Pediatric Neurology Office of Vali-e-Asr Hospital, Tehran, Iran up to 2011. Hearing loss was confirmed using Auditory Brainstem Response screening test (ABR).

**Results:** The prevalence of mildly and moderately hearing loss in neonates was determined as 3.6%. The most significant risk factors for hearing loss in neonates were neonatal icterus associated with phototherapy, respiratory distress syndrome (RDS) and lower Apgar score.

**Conclusion:** It seems to quantitative auditory system screening using ABR is necessary for all neonates; because rehabilitation support such as speech therapy and hearing training in this age period is more effective than older ages.

**Khairy MA, Abuelhamed WA, Ahmed RS, El Fouly HES, Elhawary IM.**

**[Hearing loss among high-risk newborns admitted to a tertiary Neonatal Intensive Care Unit. J Matern Fetal Neonatal Med.](#)**

2018 Jul;31(13):1756-1761. doi: 10.1080/14767058.2017.1326902. Epub 2017 May 22.

**PURPOSE:** The aim of this work is to identify the most significant risk factors for hearing impairment in high risk neonates hospitalized at our Neonatal Intensive Care Unit (NICU) and to assess the sensitivity of hearing screening tests.

**METHODS:** This study involved 260 neonates admitted to a tertiary NICU; they were classified into two groups; 150 preterm and 110 full terms with risk factors for hearing loss. The hearing screening tests performed were transient evoked otoacoustic emissions (TEOAEs) and the automated auditory brainstem response (AABR).

**RESULTS:** Forty-eight preterm neonates (32%) and 30 full term neonates (27.3%) had pathological AABR. In preterm group, mechanical ventilation more than five days, sepsis, usage of aminoglycosides, loop diuretics, vancomycin alone or in combination with aminoglycosides and prolonged duration of admission were considered risk factors of hearing affection whereas in full term group mechanical ventilation more than five days was the risk factor of hearing affection ( $p < .05$ ).

**CONCLUSIONS:** The prevalence of hearing loss is highest among high risk neonates and TEOAE and AABR were found to be reliable screening tools. Use of ototoxic drugs and mechanical ventilation for more than five days were significant risk factors for hearing loss in our study population.



Khan NB, Joseph L, Adhikari M.

[The hearing screening experiences and practices of primary health care nurses: Indications for referral based on high-risk factors and community views about hearing loss. Afr J Prim Health Care Fam Med.](#)

2018 Oct 10;10(1):e1-e11. doi: 10.4102/phcfm.v10i1.1848.

**BACKGROUND:** In South Africa, primary health care is the first point of contact with the health system for at least 85% of the population, yet early hearing detection and intervention continues to be elusive in these settings. Nurses at community level may, therefore, be missing an opportunity to identify prelingual infants with hearing losses and alter their developmental trajectory.

**AIM:** To determine primary health care nurses' experiences, practices and beliefs regarding hearing loss in infants.

**SETTING:** The study was conducted in the eThekweni District of KwaZulu-Natal, South Africa.

**METHODS:** A descriptive survey was used with quantitative methods of analysis. Fourteen primary health care clinics from the eThekweni district were selected, from which 75 nurses participated by completing a self-administered questionnaire.

**RESULTS:** At least one-third of primary health care nurses had never screened a child for hearing loss, and most clinics did not have access to basic hearing screening equipment or materials. Only 49% of nurses had access to an otoscope, while 31% used the Road to Health Development screener to check for hearing loss. None of the clinics had access to an otoacoustic emission screener nor the Swart questionnaire. Although nurses reported that they would refer to audiology services for some of the risk factors, as indicated on the Joint Committee on Infant Hearing (JCIH) 2007 list, they were less likely to refer if the child was in a neonatal intensive care unit (ICU) longer than five days, had neurodegenerative disorders, meningitis, hyperbilirubinaemia requiring blood transfusion or were undergoing chemotherapy. Less than a third of nurses always referred if the child displayed additional non-JCIH risk factors or those pertinent to the South African context. Approximately 38% reported that communities believed that hearing loss could be because of some form of spiritual or supernatural causes.

**CONCLUSION:** This study demonstrates that hearing screening and referral practices at primary health care clinics need to be strengthened. Nurses need to be capacitated to conduct basic screening, make necessary referrals, provide information to caregivers and understand community beliefs about hearing loss in order to counsel caregivers appropriately and facilitate the process of early hearing detection and intervention.

Kim BJ, Han JJ, Shin SH, Kim HS, Yang HR, Choi EH, Chang MY, Lee SY, Suh MW, Koo JW, Lee JH, Choi BY, Oh SH.

[Characterization of Detailed Audiological Features of Cytomegalovirus Infection: A Composite Cohort Study from Groups with Distinct Demographics. Biomed Res Int.](#)

2018 Aug 30;2018:7087586. doi: 10.1155/2018/7087586. eCollection 2018.

Congenital cytomegalovirus (cCMV) infection is a common congenital infection that causes sensorineural hearing loss (SNHL). Despite its substantial impact on public health and cost burden, epidemiology and clinical features of CMV-related SNHL have never been reported in the Korean populations. This study investigated the detailed audiologic phenotypes of cCMV infection to see if a specific SNHL pattern is associated with a particular clinical setting. A total of 38 patients with cCMV infection were studied retrospectively. Patients were classified into three groups with distinct demographics: clinically driven diagnosis (n=17), routine newborn CMV screening according to the NICU protocols (n=10), or referral to ENT for cochlear implant (CI) (n=11). The incidence of cCMV infection was 3.6%, showing 33.3% of SNHL among cCMV patients, 38% of asymmetric hearing loss, 29% of late-onset hearing loss, and diverse severity spectrum in patients with CMV-related SNHL. CI recipients with CMV-related SNHL showed a significantly improved speech perception. Surprisingly, in 36.4% of CI implantees, initial audiological manifestation was significant asymmetry of hearing thresholds between both ears, with better ear retaining significant residual hearing up to 50dB. CMV turns out to be a significant etiology of SNHL, first to date reported in the Korean pediatric population. Analysis of audiologic phenotypes showed a very wide spectrum of SNHL and favorable CI outcomes in case of profound deafness. Especially for the patients with asymmetric hearing loss, close surveillance of hearing should be warranted and CI could be considered on the worse side first, based on the observation of rapid progression to profound deafness of better side.

Kobas M, Bickle Graz M, Truttmann AC, Giannoni E, Meylan P, Asner SA.

[Clinical characteristics, audiological and neurodevelopmental outcomes of newborns with congenital cytomegalovirus infection. Swiss Med Wkly.](#)

2018 Jun 12;148:w14627. doi: 10.4414/smw.2018.14627. eCollection 2018.

**BACKGROUND:** Congenital cytomegalovirus (cCMV) infections are the leading nongenetic cause of congenital sensorineural hearing loss (SNHL); however the true impact of cCMV infections remains unknown.

**AIMS OF THE STUDY:** (1) To identify the number of asymptomatic and symptomatic cCMV infections diagnosed between 1999 and 2014 at the Lausanne University Hospital; (2) to describe the audiological and neurodevelopmental outcomes of infants with cCMV infection; and (3) to compare clinical outcomes between infants born to mothers with primary versus nonprimary infection.

**METHODS:** This was a single-centre, observational, exploratory, retrospective study of newborns diagnosed with cCMV infection at the Lausanne University Hospital between 1999 and 2014.

**RESULTS:** Fifty newborns with cCMV infection were identified; 39 (78%) were symptomatic at birth, of whom 29 (74%) were neurologically symptomatic. Twelve children (24%) presented with subsequent abnormal audiological and/or neurodevelopmental outcomes. Newborns born to mothers with a nonprimary infection were more often symptomatic at birth than those born to mothers with a primary infection.

**CONCLUSIONS:** All infants with subsequent SNHL or abnormal neurodevelopment were symptomatic at birth. Similar long-term neurodevelopmental and audiological outcomes were observed in infants born to mothers with a primary and nonprimary infection.

**Labaeka AA, Tongo OO, Ogunbosi BO, Fasunla JA.**

[Prevalence of Hearing Impairment Among High-Risk Newborns in Ibadan, Nigeria. \*Front Pediatr.\*](#)

2018 Jul 16;6:194. doi: 10.3389/fped.2018.00194. eCollection 2018.

The burden of severe hearing impairment is increasing with two-thirds of these hearing impaired people residing in developing countries. Newborn hearing screening helps to identify early, babies who need intervention in order to prevent future disability. Neither universal nor targeted hearing screening programme is available in Nigeria. **Objectives:** This study was carried out to assess the prevalence of hearing impairment among high-risk newborns in UCH and the associated risk factors. **Materials and Methods:** Two hundred one newborns in the neonatal unit of UCH with risk factors for hearing impairment had hearing screening done using automated auditory brainstem response (AABR) at 30, 45, and 70 dB at admission and discharge, and those that failed screening at discharge were rescreened at 6 weeks post-discharge. **Results:** Eighty-three (41.3%) and 32 (15.9%) high-risk newborns failed at admission and discharge screening respectively, and 19 (9.5%) still failed at follow up screening. The majority of hearing loss at follow up was bilateral (94.7%) and severe (52.6%). The risk factors associated with persistent hearing loss at follow up were acute bilirubin encephalopathy (RR = 11.2, CI: 1.4-90.6), IVH (RR = 8.8, CI: 1.1-71.8), meningitis (RR = 4.8, CI: 1.01-29), recurrent apnoea (RR = 2.7, CI: 1.01-7.3), severe perinatal asphyxia NNE III (RR = 7, CI: 2.4-20.2). **Conclusion:** Severe and bilateral hearing impairment is a common complication among high risk newborns in UCH persisting till 6 weeks post-neonatal care. Severe perinatal asphyxia with NNE III, ABE, IVH, meningitis and administration of amikacin for more than 5 days were significant risk factors. We recommend that SCBU graduates with these risk factors should have mandatory audiologic evaluation at discharge.

**Lantos PM, Maradiaga-Panayotti G, Barber X, Raynor E, Tucci D, Hoffman K, Permar SR, Jackson P, Hughes BL, Kind A.**

[Geographic and Racial Disparities in Infant Hearing Loss. \*Otolaryngol Head Neck Surg.\*](#)

2018 Oct 9:194599818803305. doi: 10.1177/0194599818803305. [Epub ahead of print]

**Objective** Approximately 1 to 2 of every 1000 American newborns has hearing loss identified by newborn screening. This study was designed to determine if infant hearing loss is more common in socioeconomically disadvantaged communities. **Study Design** In this retrospective study, we analyzed electronic medical record data using geostatistical models. **Setting** Infants were residents of Durham County, North Carolina, born in 2 hospitals of the Duke University Health System. This county includes the city of Durham and surrounding suburban and rural communities. **Subjects and Methods** Subjects were hearing-screened newborns, born between 2005 and 2016, whose residential address was in Durham County, North Carolina. This was a retrospective study using medical record data. We used Bayesian regression models with smoothing of coordinate date to identify both spatial and nonspatial predictors of infant hearing loss. **Results** We identified 19,348 infants from Durham County, of whom 675 had failed initial hearing screening and 191 had hearing loss confirmed on follow-up. Hearing loss was significantly associated with minority race (odds ratio [OR], 2.45; 95% confidence interval, 1.97-3.06), as well as lower gestational age and maternal sexually transmitted infections. We identified significant geographic heterogeneity, with a higher probability of hearing loss in poorer urban neighborhoods (local OR range, 0.59-1.39). Neighborhood disadvantage was a significant predictor of hearing loss, as was high local seroprevalence of cytomegalovirus (CMV) among pregnant women. **Conclusions** Urban, low-income neighborhoods have a high prevalence of infant hearing loss compared with more affluent surrounding communities, particularly among minorities. This distribution may be attributable to congenital CMV infection.

**Levit Y, Mandel D, Matot I.**

[Frequency-specific auditory brainstem response testing with age-appropriate sedation. \*Int J Pediatr Otorhinolaryngol.\*](#)

2018 May;108:73-79. doi: 10.1016/j.ijporl.2018.02.028. Epub 2018 Feb 17.

**OBJECTIVE:** Auditory brainstem response (ABR) testing is the gold-standard procedure for hearing evaluation in pediatric patients who cannot complete a behavioral hearing test. The amount of audiological information obtained depends on the quality of the patient's sleep during the test. In this retrospective database review, we aimed to assess the amount and the characteristics of the audiological information obtained in ABR testing in pediatric patients with age-appropriate sedation.

**METHODS:** A retrospective chart review was conducted on 501 consecutive ABR sedation sessions performed between January 2014 and June 2016 at the Tel Aviv Medical Center. Oral triclofos was used for the sedation of younger patients (3-24 months) and intravenous propofol for older patients (>24 months). The dataset included 370 triclofos sessions (in 337 patients) and 131 propofol sessions (in 126 patients).

**RESULTS:** None of the children developed complications, and all were discharged on the same day of the evaluation. Among the hearing-impaired children, a mean of 10 (1.8 SD) ABR threshold measurements was obtained from propofol-sedated patients and 9.4 (2.8 SD) measurements from those sedated with triclofos (P = 0.039). The major characteristics of the hearing loss, including its degree, type, and configuration, were obtained from all propofol-sedated patients and from 95% of those sedated with triclofos.

**CONCLUSIONS:** A comprehensive evaluation of hearing status can be obtained in ABR testing with age-appropriate sedation. An average number of ~10 threshold measurements were obtained during ABR testing with age-appropriate sedation, thus allowing for the evaluation of the degree, type and configuration of the hearing loss.

Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi, Mei L, Chen XP, Yang J.

[The study of detection and etiology of unilateral deafness in children.](#)

2018 Aug;32(16):1252-1254. doi: 10.13201/j.issn.1001-1781.2018.16.010.[Article in Chinese]

**Objective:**To explore the detection approach and aetiology of single-side deafness(SSD) in children and provide evidence for diagnoses and treatment. **Method:**A retrospective study was performed. Medical history and radiological data of children with unilateral deafness were analyzed. **Result:**The mean age at diagnosis was 6 years old. The mean age at diagnosis was 7.7 month because of failure of universal newborn hearing screening. Inner ear malformations were identified in 80.6% of cases. Cochlear nerve deficiency(CND) was observed in almost 77.8% of our cases, accounts for 96.6% in children with inner ear malformations. **Conclusion:**The children with SSD could be early detected by UNHS. CND is the main aetiology for SSD. MRI should be the first image choice when SSD children was diagnosed.

Lu CY, Tsao PN, Ke YY, Lin YH, Lin YH, Hung CC, Su YN, Hsu WC, Hsieh WS, Huang LM, Wu CC, Hsu CJ.

[Concurrent Hearing, Genetic, and Cytomegalovirus Screening in Newborns, Taiwan. J Pediatr.](#)

2018 Aug;199:144-150.e1. doi: 10.1016/j.jpeds.2018.02.064. Epub 2018 Apr 19.

**OBJECTIVE:** To evaluate the feasibility and potential benefits of incorporating genetic and cytomegalovirus (CMV) screenings into the current newborn hearing screening (NHS) programs.

**STUDY DESIGN:** Newborns were recruited prospectively from a tertiary hospital and a maternity clinic between May 2016 and December 2016 and were subjected to hearing screening, CMV screening, and genetic screening for 4 common mutations in deafness genes (p.V37I and c.235delC of GJB2 gene, c.919-2A>G of SLC26A4 gene, and the mitochondrial m.1555A>G). Infants with homozygous nuclear mutations or homoplasmic/heteroplasmic mitochondrial mutation (referred to as “conclusively positive genotypes”) and those who tested positive for CMV received diagnostic audiologic evaluations.

**RESULTS:** Of the total 1716 newborns enrolled, we identified 20 (1.2%) newborns with conclusively positive genotypes on genetic screening, comprising 15 newborns (0.9%) with GJB2 p.V37I/p.V37I and 5 newborns (0.3%) with m.1555A>G. Three (0.2%) newborns tested positive on CMV screening. Twelve of the 20 newborns (60%) with conclusively positive genotypes and all 3 newborns who tested positive for CMV (100%) passed NHS at birth. Diagnostic audiologic evaluations conducted at 3 months confirmed hearing impairment in 6 of the 20 infants (30%) with conclusively positive genotypes.

**CONCLUSIONS:** This study confirms the feasibility of performing hearing, genetic, and CMV screenings concurrently in newborns and provides evidence that the incorporation of these screening tests could potentially identify an additional subgroup of infants with impaired hearing that might not be detected by the NHS programs.

Mahmoudian S, Aminrasouli N, Ahmadi ZZ, Lenarz T, Farhadi M.

[Acoustic Analysis of Crying Signal in Infants with Disabling Hearing Impairment. J Voice.](#)

2018 Jul 25. pii: S0892-1997(18)30030-4. doi: 10.1016/j.jvoice.2018.05.016. [Epub ahead of print]

**OBJECTIVE:** Crying is a multimodal, dynamic behavior and the first way to communicate. Early identification of hearing impairment is critical for prevention of speech and language disorders. The present study aimed to assess the acoustic features of infant's cry signals to find possible differences between two groups including hearing-impaired (HI) infants and normal hearing (NH) control.

**METHODS:** The data were collected from 34 (17 HI, 17 NH) infants under 2 months of age. Recording of the infant cry signals was collected during the examination of the Babinski reflex and was subsequently submitted for acoustic analysis. The total duration of the recording for each infant was approximately 30 seconds. The acoustical features included fundamental frequency ( $F_0$ ), formants ( $F_1$ ,  $F_2$ , and  $F_3$ ), intensity, jitter, shimmer, ratios of  $F_2/F_1$  and  $F_3/F_1$ , ratio of harmonic to noise, and voice break. The recording device was an Olympus ws-321M voice recorder with 44,100 Hz sampling frequency in the stereo form. Praat analysis software (version 27, 3, 5) was used to analyze the crying signals. The data were then statistically analyzed using SPSS version 21.

**RESULTS:** Acoustic analysis of the crying signals showed that HI infants have lower intensity and higher  $F_0$  and voice break than NH infants. However, the other differences were not statistically significant.

**CONCLUSION:** The results of the present study demonstrated that the acoustic components including  $F_0$ , intensity, and voice break may be used as indices to discriminate HI infants from NH infants under 2 months of age. These findings can be increased our knowledge concerning the functional mechanisms of the vocal organ in HI and NH infants.

Maluleke NP, Khoza-Shangase K, Kanji A.

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

2018 Aug 1:1367493518788477. doi: 10.1177/1367493518788477. [Epub ahead of print]

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

Melo RS, Lemos A, Raposo MCF, Belian RB, Ferraz KM.

[Balance performance of children and adolescents with sensorineural hearing loss: Repercussions of hearing loss degrees and etiological factors. Int J Pediatr Otorhinolaryngol.](#)

2018 Jul;110:16-21. doi: 10.1016/j.ijporl.2018.04.016. Epub 2018 Apr 21.

**INTRODUCTION:** Some studies have demonstrated a parallelism between the extent of hearing loss and the frequency of vestibular dysfunction in children with sensorineural hearing loss (SNHL). Despite this, little is known about the repercussion of degrees of hearing loss and etiological factors on the balance performance in this children.

**OBJECTIVE:** Compare the balance performance between normal hearing (NH) children and those with SNHL, considering the sex and age range of the sample, and analyze balance performance according to the degrees of hearing loss and etiological factors in the latter group.

**METHODS:** Cross-sectional study that assessed 96 children (48 NH and 48 with SNHL), aged between 7 and 18 years old. The balance performance was assessed by the Brazilian version of the Pediatric Balance Scale, validated for Brazilian child population and the Mann-Whitney test used for statistical analysis.

**RESULTS:** The group with SNHL showed lower average balance performance compared to NH ( $p = 0.000$ ). This was also observed when the children were grouped by sex: female and male ( $p = 0.001$ ). The same difference occurred when the children were stratified by age group: 7-14 years old ( $p = 0.000$ ). There were no differences between the balance performance of the groups according to the degrees of hearing loss ( $p = 0.236$ ) and the children with prematurity or post-natal meningitis as an etiological factor demonstrated the worst balance performance.

**CONCLUSION:** The children with SNHL showed worse balance performance compared to NH of the same sex and age range between seven to fourteen years. There were no differences between balance performance and hearing loss degrees, and those children with prematurity or post-natal meningitis as an etiological factor demonstrated the worst balance performances.

Musacchia G, Ortiz-Mantilla S, Roesler CP, Rajendran S, Morgan-Byrne J, Benasich AA.

[Effects of noise and age on the infant brainstem response to speech. Clin Neurophysiol.](#)

2018 Aug 30. pii: S1388-2457(18)31191-X. doi: 10.1016/j.clinph.2018.08.005. [Epub ahead of print]

**OBJECTIVE:** Background noise makes hearing speech difficult for people of all ages. This difficulty can be exacerbated by co-occurring developmental deficits that often emerge in childhood. Sentence-type speech-in-noise (SIN) tests are available clinically but cannot be administered to very young individuals. Our objective was to examine the use of an electrophysiological test of SIN, suitable for infants, to track developmental trajectories.

**METHODS:** Speech-evoked brainstem potentials were recorded from 30 typically-developing infants in quiet and +10 dB SNR background noise. Infants were divided into two age groups (7-12 and 18-24 months) and examined across development. Spectral power of the frequency following response (FFR) was computed using a fast Fourier Transform. Cross-correlations between quiet and noise responses were computed to measure encoding resistance to noise.

**RESULTS:** Older infants had more robust FFR encoding in noise and had higher quiet-noise correlations than their younger counterparts. No group differences were observed in the quiet condition.

**CONCLUSIONS:** By two years of age, infants show less vulnerability to the disruptive effects of background noise, compared to infants under 12 months.

**SIGNIFICANCE:** Speech-in-noise electrophysiology can be easily recorded across infancy and provides unique insights into developmental differences that tests conducted in quiet may miss.

Netten AP, Rieffe C, Ketelaar L, Soede W, Gadow KD, Frijns JHM.

[Terrible Twos or Early Signs of Psychopathology? Developmental Patterns in Early Identified Preschoolers With Cochlear Implants Compared With Hearing Controls. Ear Hear.](#)

2018 May/ Jun;39(3):495-502. doi: 10.1097/AUD.0000000000000500.

**OBJECTIVE:** Cochlear implants (CIs) have dramatically improved the lives of children who are deaf or hard of hearing; however, little is known about its implications for preventing the development of psychiatric symptoms in this at-risk population. This is the first longitudinal study to examine the early manifestation of emotional and behavioral disorders and associated risk and protective factors in early identified preschoolers with CIs compared with hearing peers.

**DESIGN:** Participants were 74 children with CIs and 190 hearing controls between ages 1 and 5 years (mean age, 3.8 years). Hearing loss was detected using the Newborn Hearing Screening in The Netherlands and Flanders. Parents completed the Early Childhood Inventory-4, a well-validated measure, to evaluate the symptoms of DSM-IV-defined psychiatric disorders, during three consecutive years. Language scores were derived from each child's medical notes.

**RESULTS:** Children with CIs and hearing controls evidenced comparable levels of disruptive behavior and anxiety/depression (which increased with age in both groups). Greater proficiency in language skills was associated with lower levels of psychopathology. Early CI and longer duration of CI use resulted in better language development. In turn, higher early language skills served as a protective factor against the development of disruptive behavior symptoms.

**CONCLUSIONS:** This longitudinal study uniquely shows that improvement in language skills mitigates the development of early signs of psychopathology. Early identification of hearing loss and CIs help children improve their language skills.

Parab SR, Khan MM, Kulkarni S, Ghaisas V, Kulkarni P.

[Neonatal Screening for Prevalence of Hearing Impairment in Rural Areas. Indian J Otolaryngol Head Neck Surg.](#)

2018 Sep;70(3):380-386. doi: 10.1007/s12070-018-1386-4. Epub 2018 May 4.

Hearing is one of the most important sense organs for man. Hearing loss is often associated with delayed speech and language development in young children. Early identification and intervention improves the chance a child gets to lesser delays in



development and improving the overall quality of life. To find out the prevalence of hearing loss in neonates in the rural taluka of Maval, Pune, Maharashtra, India. Prospective Non Randomized Clinical Study. The study was carried out between April 2012 and April 2015. A total of 8192 babies were screened across various centers around the Maval area. The babies who had some high risk factors were 1683 in number and babies who had no high risk factors i.e. well babies were 6509. In our study, the overall prevalence of hearing loss in neonates in Maval taluka of Maharashtra was found to be 3.54 per 1000 live births, in normal born neonates (well babies) was 1.689 per 1000 births, in high risk babies was 10.69 per 1000 high risk births. The prevalence of low birth weight neonates, hyperbilirubinemia neonates and neonates with craniofacial abnormalities developing hearing impairment was found to be 5.9, 3.56 and 1.18 per 1000 high risk births respectively. India is the second most populated country in the world with nearly a fifth of the world's population. There is a need for the universal neonatal screening for deafness for earlier detection of deafness and rehabilitation.

**Ia Paz EMC, Pedro M, Yarza TKL, Lagrana-Villagracia SM, Amoranto AJP, Jover EJM, Domine MTB, Chiong CM, Santos-Cortez RLP.**

[Genetic counseling in an indigenous Filipino community with a high prevalence of A2ML1-related otitis media. J Community Genet.](#)

2018 Jun 15. doi: 10.1007/s12687-018-0368-2. [Epub ahead of print]

In this report, we describe the knowledge and beliefs on causes and management of otitis media of an indigenous Filipino community with a high prevalence of otitis media that is associated with an A2ML1 variant. Community lectures and individual genetic counseling were provided as intervention. Knowledge, beliefs, and health care-seeking behavior pertaining to otitis media were assessed pre- and post-genetic counseling. Twenty-five heads of households were interviewed. Beliefs regarding etiology of ear discharge varied widely, with swimming in the sea as the most commonly cited cause of ear discharge. During the post-counseling session, poor personal hygiene, dirty environment, and familial inheritance were mentioned as risk factors for otitis media or ear discharge. Knowledge about the genotypes for the A2ML1 variant and otitis media diagnoses within the household influenced beliefs on the role of hygiene and genetic susceptibility to otitis media and attitudes towards health care-seeking behavior. Genetic counseling was associated with variable improvement in knowledge on otitis media and in their understanding of genetic susceptibility to otitis media due to the A2ML1 variant. Language barriers, literacy level, cultural factors, and the complex nature of genetic information made genetic counseling in the particular population a challenge. Insights drawn from this experience recommend follow-up visits in the community.

**Percy-Smith L, Hallstrøm M, Josvassen JL, Mikkelsen JH, Nissen L, Dieleman E, Cayé-Thomasen P.**

[Differences and similarities in early vocabulary development between children with hearing aids and children with cochlear implant enrolled in 3-year auditory verbal intervention. Int J Pediatr Otorhinolaryngol.](#)

2018 May;108:67-72. doi: 10.1016/j.ijporl.2018.02.030. Epub 2018 Feb 21

**OBJECTIVE:** The overall objective of this study was to evaluate the implementation of a Nordic Auditory Verbal (AV) intervention for children with all degrees and types of hearing impairment (HI) using all kinds of hearing technology. A first specific objective was to identify differences and similarities in early vocabulary development between children with cochlear implant (CI) compared with children with hearing aids (HAs)/Bone anchored hearing aids (Bahs) enrolled in a 3-year AV program, and to compare the group of children with HI to a control group of children with normal hearing (NH). A second specific objective was to study universal neonatal hearing screening (UNHS) using the 1-3-6 Early Hearing Detection and Intervention (EHDI) guidelines.

**INTRODUCTION:** Effect of AV intervention for children with HI using different hearing technology is not thoroughly studied. It is relevant to question, whether children with mild to moderate HI encounter the same intensive need for AV intervention as children with congenital deafness.

**METHODS:** A longitudinal and comparative study design was used involving two cohorts of children, i.e. 36 children with CI and 19 children with HA/Bahs. The children were the first in Denmark to receive a 3-year AV intervention by formally trained AV-practitioners. Children were tested annually with standardized speech and language tests, i.e. Peabody Picture Vocabulary test, Reynell test and a Danish test for active vocabulary, Viborgmateriale. Categorical variables were compared using Fischer's exact test and continuous variables were compared using Wilcoxon-Mann-Whitney test, as data was not normally distributed.

**RESULTS:** Median age of diagnosis was 6 months and median age at intervention was 13 and 12 months respectively. There was no statistically significant difference between the two groups in terms of scores according to age equivalency for the three tests. However, there was a significant difference between children with HI regardless of hearing technology and children with NH.

**CONCLUSION:** Children with HI progressed over a three-year period, but they did not reach the same level as children with NH. The high completion rate of 98,2% of families over a three-year period indicates the relevance of AV practice in a Nordic country. Children were diagnosed later than 3 months and intervention also started later than recommended. A result that warrants further investigation.

**Rahimi V, Mohammadkhani G, Javadi F.**

[Improving universal newborn hearing screening outcomes by conducting it with thyroid screening. Int J Pediatr Otorhinolaryngol.](#)

2018 Aug;111:111-114. doi: 10.1016/j.ijporl.2018.06.002. Epub 2018 Jun 5.

**OBJECTIVES:** One of the most important factors that can improve hearing screening indicators is testing infants after 48 h of birth. The neonatal thyroid screening program is done during the third to fifth day after birth in many countries. So this screening is done at the appropriate time for hearing screening. The aim of the present study was to evaluate hearing screen-

ing outcomes (the referral rate, false positive rate, and positive predictive value) conducted with the thyroid screening at the healthcare centers and compare the results with hospital before discharge the infant.

#### **METHODS:**

This was a prospective exploratory cohort study. The study population included all the newborns at a hospital (group 1) and newborns who were referred to healthcare centers for thyroid screening (group 2), except for infants with risk factors, from March 2012 to December 2017. Transient evoked otoacoustic emissions (TEOAE) and automatic auditory brainstem response (AABR) were used for the evaluation. The results were compared between the two groups.

**RESULTS:** Of the 4729 newborns, who participated in the study, 3001 were referred from a hospital (group 1) and 1728 from two healthcare centers (group 2). The referral rate in group 1 and 2 was 16.1% and 7.6%, respectively. Also, the false positive rate in group 1 and 2 was 15.9% and 7.6%, respectively. Our study showed that the referral rate and false positive rate of hearing screening in group 2 were significantly lower than that in group 1 ( $p < 0.001$ ). The positive predictive value in group 1 was significantly higher than that in group 2 ( $p < 0.05$ ). There was no significant sex difference in any of the variables.

**CONCLUSIONS:** Our results showed that performing the hearing screening during the thyroid program, instead of the hospital could be significantly improved screening outcomes and suggest that hearing and thyroid screening together after discharge from the hospital could be a good opportunity to introduce new framework for hearing screening in many countries.

**Rechia IC, Fattore IM, Moraes AB, Biaggio EPV, Souza APR.**

**[Auditory maturation and psychological risk in the first year of life. \*Codas\*.](#)**

2018 Jul 19;30(4):e20170142. doi: 10.1590/2317-1782/20182017142.

**PURPOSE:** To assess the potential association between psychological risk and limited auditory pathway maturation.

**METHODS:** In this longitudinal cohort study, 54 infants (31 non-risk and 23 at-risk) were assessed from age 1 to 12 months. All had normal hearing and underwent assessment of auditory maturation through cortical auditory evoked potentials testing. Psychological risk was assessed with the Child Development Risk Indicators (CDRIs) and PREAUT signs. A variety of statistical methods were used for analysis of results.

**RESULTS:** Analysis of P1 and N1 latencies showed that responses were similar in the both groups. Statistically significant differences between-groups were observed only for the variables N1 latency and amplitude at 1 month. Significant maturation occurred in both groups ( $p < 0.05$ ). There was moderate correlation between P1 latency and Phase II CDRIs, which demonstrates that children with longer latencies at age 12 months were more likely to exhibit absence of these indicators in Phase II and, therefore, were at greater psychological risk. The Phase II CDRIs also correlated moderately with P1 and N1 latencies at 6 months and N1 latencies at 1 month; again, children with longer latency were at increased risk.

**CONCLUSION:** Less auditory pathway maturation correlated with presence of psychological risk. Problems in the mother-infant relationship during the first 6 months of life are detrimental not only to cognitive development, but also to hearing. A fragile relationship may reflect decreased auditory and linguistic stimulation.

**Rice ME, Galang RR, Roth NM, Ellington SR, Moore CA, Valencia-Prado M, Ellis EM, Tufa AJ, Taulung LA, Alfred JM, Pérez-Padilla J, Delgado-López CA, Zaki SR, Reagan-Steiner S, Bhatnagar J, Nahabedian JF 3rd, Reynolds MR, Yeargin-Allsopp M, Viens LJ, Olson SM, Jones AM, Baez-Santiago MA, Oppong-Twene P, VanMaldeghem K, Simon EL, Moore JT, Polen KD, Hillman B, Ropeti R, Nieves-Ferrer L, Marcano-Huertas M, Masao CA, Anzures EJ, Hansen RL Jr, Pérez-Gonzalez SI, Espinet-Crespo CP, Luciano-Román M, Shapiro-Mendoza CK, Gilboa SM, Honein MA.**

**[Vital Signs: Zika-Associated Birth Defects and Neurodevelopmental Abnormalities Possibly Associated with Congenital Zika Virus Infection - U.S. Territories and Freely Associated States, 2018. \*MMWR Morb Mortal Wkly Rep\*.](#)**

2018 Aug 10;67(31):858-867. doi: 10.15585/mmwr.mm6731e1.

**INTRODUCTION:** Zika virus infection during pregnancy causes serious birth defects and might be associated with neurodevelopmental abnormalities in children. Early identification of and intervention for neurodevelopmental problems can improve cognitive, social, and behavioral functioning.

**METHODS:** Pregnancies with laboratory evidence of confirmed or possible Zika virus infection and infants resulting from these pregnancies are included in the U.S. Zika Pregnancy and Infant Registry (USZPIR) and followed through active surveillance methods. This report includes data on children aged  $\geq 1$  year born in U.S. territories and freely associated states. Receipt of reported follow-up care was assessed, and data were reviewed to identify Zika-associated birth defects and neurodevelopmental abnormalities possibly associated with congenital Zika virus infection.

**RESULTS:** Among 1,450 children of mothers with laboratory evidence of confirmed or possible Zika virus infection during pregnancy and with reported follow-up care, 76% had developmental screening or evaluation, 60% had postnatal neuroimaging, 48% had automated auditory brainstem response-based hearing screen or evaluation, and 36% had an ophthalmologic evaluation. Among evaluated children, 6% had at least one Zika-associated birth defect identified, 9% had at least one neurodevelopmental abnormality possibly associated with congenital Zika virus infection identified, and 1% had both.

**CONCLUSION:** One in seven evaluated children had a Zika-associated birth defect, a neurodevelopmental abnormality possibly associated with congenital Zika virus infection, or both reported to the USZPIR. Given that most children did not have evidence of all recommended evaluations, additional anomalies might not have been identified. Careful monitoring and evaluation of children born to mothers with evidence of Zika virus infection during pregnancy is essential for ensuring early detection of possible disabilities and early referral to intervention services.

Sato T, Nakazawa M, Takahashi S, Mizuno T, Sato A, Noguchi A, Sato M, Katagiri S, Yamada T.

[Leaflets and continual educational offerings led to increased coverage rate of newborn hearing screening in Akita. \*Auris Nasus Larynx\*.](#)

2018 Aug;45(4):673-679. doi: 10.1016/j.anl.2017.11.014. Epub 2017 Nov 28.

**OBJECTIVE:** Newborn hearing screening (NHS) has been actively performed in Japan since 2001. The NHS coverage rate has increased each year in Akita Prefecture. We analyzed the details of the NHS program and how the Akita leaflets and the many educational offerings about the importance of NHS led to the high NHS coverage rate.

**METHODS:** A retrospective study was conducted in liveborn newborns in hospitals and in clinics where hearing screening was performed from the program's beginning in 2001 through the end of 2015. We describe the chronological history of NHS. The outcome data of NHS were collected from our department and analyzed.

**RESULTS:** From the founding of the program in 2001 to 2015, the live birth rate in Akita continually declined. Nevertheless, the number of infants receiving NHS rose each year. Since 2012, the coverage rate of NHS has been over 90%. From 2001 to 2015, 75,331 newborns constituted the eligible population for the NHS program. Since 2012, the number of NHS tests has stabilized. We prepared educational leaflets for Akita Prefecture early in 2002. We also provided many educational classes about the importance of NHS for not only pregnant women but also professionals including obstetricians and gynecologists, pediatricians and municipal staff members. The NHS program received the complete endorsement of the Akita Association of Obstetricians and Gynecologists in 2010. The largest increase in the NHS coverage rate occurred from 2001 to 2002, and the second largest increase occurred from 2009 to 2010. The number of participating institutions increased the coverage rate. The coverage rate is strongly correlated with the number of participating institutions ( $r_s=0.843$ ,  $p<0.001$ , Spearman's rank correlation coefficient). Comparing the coverage rate for 5 years before and after the Akita Association of Obstetricians and Gynecologists reached their consensus on the importance of NHS, the coverage rate after 2010 was significantly higher than before 2010 ( $p<0.001$ , paired sample t-test).

**CONCLUSION:** The NHS coverage rate ultimately reached 95.4% without need for legislation or subsidization. The number of participating institutions increased each year, and the number of NHS tests and the coverage rate increased proportionately. The number of participating institutions statistically has a strong correlation with the number of NHS tests and the coverage rate. Our research indicates that the Akita leaflets and the provision of educational sessions about the importance of NHS were the most significant factors in establishing the high NHS coverage rate.

Shim YJ, Choi BY, Park KH, Lee H, Jung YM, Kim YM.

[Inflammatory and Immune Proteins in Umbilical Cord Blood: Association with Hearing Screening Test Failure in Preterm Neonates. \*Mediators Inflamm\*.](#)

2018 Sep 19;2018:4209359. doi: 10.1155/2018/4209359. eCollection 2018.

**Objective:** We aimed to determine whether elevated levels of various inflammatory and immune proteins in umbilical cord blood are associated with an increased risk of newborn hearing screening (NHS) test failure in preterm neonates.

**Methods:** This retrospective cohort study included 127 premature singleton infants who were born at  $\leq 33.6$  weeks. Umbilical cord plasma at birth was assayed for interleukin (IL)-6, complement C3a and C5a, matrix metalloproteinase (MMP)-9, macrophage colony-stimulating factor (M-CSF), and endostatin levels using ELISA kits. Neonatal blood C-reactive protein (CRP) levels were measured within 2 hours of birth. The primary outcome measure was a uni- or bilateral refer result on an NHS test. Univariate and multivariate analyses were applied.

**Results:** Fifteen (11.8%) infants failed the NHS test. In the univariate analyses, high IL-6 and low C3a levels in umbilical cord plasma, funisitis, and an elevated CRP level ( $>5$  mg/L) in the immediate postnatal period were significantly associated with NHS test failure. However, the levels of umbilical cord plasma MMP-9, C5a, M-CSF, and endostatin were not significantly different between infants who passed and those who failed the NHS test. Multiple logistic regression analyses indicated that elevated umbilical cord plasma C3a levels were independently associated with a reduced risk of NHS test failure, whereas elevated levels of umbilical cord plasma IL-6 and high CRP levels in the immediate postnatal period were significantly associated with NHS test failure.

**Conclusions:** Our data demonstrated that in preterm neonates, a systemic fetal inflammatory response reflected by umbilical cord plasma IL-6 and immediate postnatal CRP levels may contribute to the risk for NHS test failure, whereas the changes in complement activation fragments initiated *in utero* may have protective effect of hearing screen failure.

da Silva LS, Ribeiro GE, Montovani JC, Silva DPCD.

[The effect of peri-intraventricular hemorrhage on the auditory pathway of infants. \*Int J Pediatr Otorhinolaryngol\*.](#)

2018 Sep;112:24-26. doi: 10.1016/j.ijporl.2018.06.026. Epub 2018 Jun 13.

**OBJECTIVE:** To verify the effect of peri-intraventricular hemorrhage on the auditory pathway of preterm infants.

**METHOD:** It is a non-concurrent cohort study. This study was conducted in a tertiary public. Preterm infants with peri-intraventricular hemorrhage comprised the study group, and preterm infants without peri-intraventricular hemorrhage were included as a comparison group, both were similar in relation to gestational age and risk indicators for hearing loss. Participants had to meet the following inclusion criteria: have been born at the study site, presence of otoacoustic emissions by transient stimulus in both ears and brainstem auditory evoked potentials with all components bilaterally identified.

**RESULTS:** 44 infants with an average age of 3 months with peri-intraventricular hemorrhage and 2,6 months without peri-intraventricular hemorrhage met the inclusion criteria. Regarding the brainstem auditory evoked potentials results, a significant increase was observed in absolute latency values of waves I, III and V, as well as in the interpeak intervals I-III and I-V, bilaterally, in infants with peri-intraventricular hemorrhage.

**CONCLUSION:** This study concluded that infants with peri-intraventricular hemorrhage presented a delay in the neural conduction of sound, which justifies the monitoring of the auditory function in these infants during the period of language development.

Sunwoo W, Lee WW, Choi BY.

[Extremely common radiographic finding of cochlear nerve deficiency among infants with prelingual single-sided deafness and its clinical implications. \*Int J Pediatr Otorhinolaryngol.\*](#)

2018 Sep;112:176-181. doi: 10.1016/j.ijporl.2018.07.013. Epub 2018 Jul 10.

**OBJECTIVES:** To clarify the common radiographic findings of audiotically documented prelingual single-sided deafness (SSD) and identify the prevalence of cochlear nerve deficiency (CND) in SSD infants referred from the newborn hearing screening program.

**METHODS:** Between March 2012 and March 2017, the records of all infants referred to our otology clinic after undergoing newborn hearing screening program were retrospectively reviewed. Twenty-four consecutive well infants without risk factors who had a confirmed diagnosis of prelingual SSD under the age of 1 year and who underwent internal auditory canal (IAC) magnetic resonance imaging (MRI) were included. The sizes of cochlear nerve (CN), IAC, and cochlear nerve canal (CNC) were measured on MRI. The presence of CND was visually determined by comparing the CN size to the ipsilateral facial nerve (FN) in the affected side via an oblique sagittal view of IAC MRI and defined when CN was absent or smaller than FN.

**RESULTS:** CND was seen in all 24 deaf ears (100%) on MRI. There was one with incomplete partition type I, and another with combined cochleovestibular nerve absence. Twenty-four subjects demonstrated either an absent (20/24, 83.3%) or small (4/24, 16.7%) CN. When the absent and small CN groups were compared, the former group had a higher prevalence of narrow CNC and narrow IAC. Of the 20 infants without identifiable CN on the affected side, 17 (85%) had narrow IAC and 17 (85%) had narrow CNC. In the 20 ears with absent CN, only one had both normal-sized IAC and CNC.

**CONCLUSION:** The contribution of CND to prelingual SSD in Korean infants reached 100%, according to IAC MRI alone.

Suskind DL, Leung CYY, Webber RJ, Hundertmark AC, Leffel KR, Fuenmayor Rivas IE1, Grobman WA.

[Educating Parents About Infant Language Development: A Randomized Controlled Trial. \*Clin Pediatr \(Phila\).\*](#)

*Clin Pediatr (Phila).*

2018 Jul;57(8):945-953. doi: 10.1177/0009922817737079. Epub 2017 Oct 26.

A total of 427 women (aged 18-45 years) who delivered a singleton neonate without serious medical complications were randomized to watch either an educational intervention (n = 225) or the sudden infant death syndrome (n = 202) video. Linear mixed models showed that the intervention women significantly gained knowledge over time. Knowledge gain was largest among high-socioeconomic status (high-SES) and middle-SES English-speaking, smaller among low-SES Spanish-speaking, and nonsignificant among low-SES English-speaking women. Analysis of deviance revealed that the intervention women of all SES learned strategies fostering secure attachment and language acquisition. Participants considered watching an educational video alongside the universal newborn hearing screening (UNHS) conveniently timed. The intervention women were more likely than the control women to recognize the importance of timely UNHS follow-up.

Takahashi M, Arai Y, Sakuma N, Yabuki K, Sano D, Nishimura G, Oridate N, Usami SI.

[Cochlear volume as a predictive factor for residual-hearing preservation after conventional cochlear implantation. \*Acta Otolaryngol.\*](#)

2018 Apr;138(4):345-350. doi: 10.1080/00016489.2017.1393840. Epub 2017 Nov 17.

**OBJECTIVE:** The preservation of residual hearing after conventional cochlear implantation (CI) is frequently observed when atraumatic soft surgery is adopted. The purpose of this study was to elucidate the predictive factors for residual hearing preservation after atraumatic CI.

**PATIENTS:** This study included 46 patients who underwent CI based on an atraumatic technique using a standard-length flexible electrode implant through a round window approach.

**MAIN OUTCOME MEASURE:** Cochlear volume was measured using magnetic resonance imaging (MRI). Cochlear duct length (CDL) was taken as the length of the scala media measured using computed tomography (CT). The association between residual hearing preservation and cochlear volume/CDL was then examined.

**RESULT:** Cochlear volume and CDL were significantly larger in patients with complete hearing preservation than in those with hearing loss. Multivariate logistic regression analysis revealed that cochlear volume was a significant predictive factor for residual hearing preservation.

**CONCLUSION:** Residual hearing preservation after conventional CI was observed in patients with a larger cochlear volume and longer CDL. Cochlear volume could be a predictive factor for residual hearing preservation after conventional CI.

Thomson V, Yoshinaga-Itano C.

[The Role of Audiologists in Assuring Follow-Up to Outpatient Screening in Early Hearing Detection and Intervention Systems. \*Am J Audiol.\*](#)

2018 Sep 12;27(3):283-293. doi: 10.1044/2018\_AJA-17-0113.

**Purpose:** The purpose of this study was to investigate the role of audiology involvement and other factors associated with failure to follow through from the initial hearing screening to the second outpatient screen.

**Method:** Linear regression, logistical regression, and descriptive analyses were used across demographic and hospital variables associated with infants who did not receive a follow-up outpatient screen.

**Results:** The results included birthing hospital outpatient rescreen rates from January 1, 2005, through December 31, 2005. Variables were collected from the birth certificate and hospital surveys. Results showed higher loss to follow-up/documentation to outpatient screen for (a) infants born in hospitals with low rates for returning for follow-up, (b) infants born in hospitals that did not have an audiologist involved, (c) infants who were Hispanic, (d) infants who were born to mothers who were not married, (e) infants with mother's with < 12 years of education, and (f) infants with Apgar scores of 7 or below.



**Conclusions:** The findings were used to identify quality improvement strategies to decrease the loss to follow-up. Strategies included ensuring audiology support, providing information in the parent's native language, educating personnel in the newborn intensive care units, developing and disseminating information in Spanish in written form, and educating hospitals on the importance of scheduling the outpatient rescreening before hospital discharge.

**Tokat T, Catli T, Bayrak F, Bozkurt EB, Olgun L.**

[Cochlear Implantation in Postmeningitic Deafness. \*J Craniofac Surg.\*](#)

2018 May;29(3):e245-e248. doi: 10.1097/SCS.0000000000004265.

**PURPOSE:** The aim of this study is to evaluate long-term outcomes of cochlear implantation (CI) in patients with postmeningitic deafness.

**METHODS:** Twenty-seven patients with severe to profound hearing loss due to bacterial meningitis and received CI were the subjects of this study. Surgical findings and long-term audiological performances were evaluated. Speech perception and speech intelligibility of the implanted patients were evaluated with the categories of auditory performance-II (CAP-II) test and speech intelligibility rating (SIR) test, respectively.

**RESULTS:** Eighteen of the 27 patients had received full electrode insertion through the patent cochlear lumen. Remaining 9 patients had varying degrees of ossification throughout the cochlea and needed to be drilled to achieve partial electrode insertion. None of the patients exhibited surgical complication. Scores in both test batteries (CAP-II and SIR) were comparable between patients who received full or partial electrode insertion ( $P>0.05$ ).

**CONCLUSION:** Cochlear implantation after postmeningitic deafness has favorable outcomes especially in long term. Although this type of inner ear pathology may require special considerations during surgery, it is a relatively safe procedure.

**Turchetta R, Conti G, Marsella P, Orlando MP, Picciotti PM, Frezza S, Russo FY, Scorpecci A, Cammeresi MG, Giannantonio S, Greco A, Ralli M.**

[Universal newborn hearing screening in the Lazio region, Italy. \*Ital J Pediatr.\*](#)

2018 Aug 24;44(1):104. doi: 10.1186/s13052-018-0534-5.

**BACKGROUND:** The introduction of Universal Newborn Hearing Screening (UNHS) programs has drastically contributed to the early diagnosis of hearing loss in children, allowing prompt intervention with significant results on speech and language development in affected children. UNHS in the Lazio region has been initially deliberated in 2012; however, the program has been performed on a universal basis only from 2015. The aim of this retrospective study is to present and discuss the preliminary results of the UNHS program in the Lazio region for the year 2016, highlighting the strengths and weaknesses of the program.

**METHODS:** Data from screening facilities in the Lazio region for year 2016 were retrospectively analyzed. Data for Level I centers were supplied by the Lazio regional offices; data for Level II and III centers were provided by units that participated to the study.

**RESULTS:** During 2016, a total of 44,805 babies were born in the Lazio region. First stage screening was performed on 41,821 children in 37 different birth centers, with a coverage rate of 93.3%. Of these, 38,977 (93.2%) obtained a "pass" response; children with a "refer" result in at least one ear were 2844 (6.8%). Data from Level II facilities are incomplete due to missing reporting, one of the key issues in Lazio UNHS. Third stage evaluation was performed on 365 children in the three level III centers of the region, allowing identification of 70 children with unilateral (40%) or bilateral (60%) hearing loss, with a prevalence of 1.6/1000.

**CONCLUSIONS:** The analysis of 2016 UNHS in the Lazio region allowed identification of several strengths and weaknesses of the initial phase of the program. The strengths include a correct spread and monitoring of UNHS among Level I facilities, with an adequate coverage rate, and the proper execution of audiological monitoring and diagnosis among Level III facilities. Weakness, instead, mainly consisted in lack of an efficient and automated central process for collecting, monitoring and reporting of data and information.

**Uddén F, Filipe M, Reimer Å, Paul M, Matuschek E, Thegerström J, Hammerschmidt S, Pelkonen T, Riesbeck K.**

[Aerobic bacteria associated with chronic suppurative otitis media in Angola. \*Infect Dis Poverty.\*](#)

2018 May 3;7(1):42. doi: 10.1186/s40249-018-0422-7.

**BACKGROUND:** Chronic suppurative otitis media (CSOM) is an important cause of hearing loss in children and constitutes a serious health problem globally with a strong association to resource-limited living conditions. Topical antibiotics combined with aural toilet is the first-hand treatment for CSOM but antimicrobial resistance and limited availability to antibiotics are obstacles in some areas. The goal of this study was to define aerobic pathogens associated with CSOM in Angola with the overall aim to provide a background for local treatment recommendations.

**METHODS:** Samples from ear discharge and the nasopharynx were collected and cultured from 152 patients with ear discharge and perforation of the tympanic membrane. Identification of bacterial species was performed with matrix-assisted laser desorption/ionization-time of flight mass spectrometry and pneumococci were serotyped using multiplex polymerase chain reactions. Antimicrobial susceptibility testing was done according to EUCAST.

**RESULTS:** One hundred eighty-four samples from ear discharge and 151 nasopharyngeal swabs were collected and yielded 534 and 289 individual isolates, respectively. In all patients, correspondence rate of isolates from 2 ears in patients with bilateral disease was 27.3% and 9.3% comparing isolates from the nasopharynx and ear discharge, respectively. *Proteus* spp. (14.7%), *Pseudomonas aeruginosa* (13.2%) and *Enterococcus* spp. (8.8%) were dominating pathogens isolated from ear discharge. A large part of the remaining species belonged to *Enterobacteriaceae* (23.5%). *Pneumococci* and *Staphylococcus aureus* were detected in approximately 10% of nasopharyngeal samples. Resistance rates to quinolones exceeded 10% among *Enterobacteriaceae* and was 30.8% in *S. aureus*, whereas 6.3% of *P. aeruginosa* were resistant.

**CONCLUSIONS:** The infection of the middle ear in CSOM is highly polymicrobial, and isolates found in nasopharynx do not correspond well with those found in ear discharge. Pathogens associated with CSOM in Angola are dominated by gram-negatives including Enterobacteriaceae and *P. aeruginosa*, while gram-positive enterococci also are common. Based on the results of antimicrobial susceptibility testing topical quinolones would be the preferred antibiotic therapy of CSOM in Angola. Topical antiseptics such as aluminium acetate, acetic acid or boric acid, however, may be more feasible options due to a possibly emerging antimicrobial resistance.

van Beeck Calkoen EA, Merkus P, Goverts ST, van de Kamp JM, Mulder MF, Sanchez Aliaga E, Hensen EF. [Evaluation of the outcome of CT and MR imaging in pediatric patients with bilateral sensorineural hearing loss. Int J Pediatr Otorhinolaryngol.](#)

2018 May;108:180-185. doi: 10.1016/j.ijporl.2018.02.022. Epub 2018 Feb 15.

**OBJECTIVE:** To evaluate the clinically relevant abnormalities as visualized on CT and MR imaging in children with symmetric and asymmetric bilateral sensorineural hearing loss (SNHL), in relation to age and the severity of hearing loss.

**STUDY DESIGN:** Retrospective cohort study.

**SETTING:** Tertiary referral otology and audiology center.

**PATIENTS AND DIAGNOSTIC INTERVENTIONS:** From January 2006 until January 2016, a total of 207 children diagnosed with symmetric and asymmetric bilateral SNHL were included. They underwent CT and/or MR imaging for the evaluation of the etiology of their hearing loss.

**MAIN OUTCOME MEASURES:** Radiologic abnormalities associated with SNHL.

**RESULTS:** 302 scans were performed in 207 children (median age of 0.8 years old) with bilateral SNHL. The most frequently identified cause of bilateral SNHL was a malformation of the labyrinth. The combined diagnostic yield of CT and MR imaging was 32%. The diagnostic yield of MR (34%) was considerably higher than that of CT (20%). We found a higher rate of abnormalities in children with profound hearing loss (41%) compared to milder hearing loss (8-29%), and in asymmetric SNHL (52%) compared to symmetric SNHL (30%).

**CONCLUSION:** Imaging is essential in the etiologic evaluation of children with bilateral SNHL. The highest diagnostic yield is found in children with bilateral asymmetric SNHL or profound SNHL. Based on our findings, MR is the primary imaging modality of choice in the etiological evaluation of children with bilateral SNHL because of its high diagnostic yield.

Waissbluth S, Del Valle Á, Chuang A, Becker A.

[Incidence and associated risk factors for platinum-induced ototoxicity in pediatric patients. Int J Pediatr Otorhinolaryngol.](#)

2018 Aug;111:174-179. doi: 10.1016/j.ijporl.2018.06.021. Epub 2018 Jun 12.

**OBJECTIVES:** Platinum-based chemotherapy is effective against a variety of pediatric malignancies. Unfortunately, the use of cisplatin and carboplatin can lead to permanent and progressive sensorineural hearing loss which can affect the quality of life of cancer survivors. The objectives of this study were to evaluate the incidence of platinum-induced ototoxicity in children and analyze potential risk factors.

**METHODS:** Prospective cohort study. All pediatric patients receiving chemotherapy with cisplatin and/or carboplatin from 01/2012 until 10/2017 were included. Hearing evaluations were performed before every chemotherapy cycle, and following the end of chemotherapy, with auditory brainstem response, otoacoustic emissions and/or audiometry. Demographics, cumulative doses, cranial irradiation and exposure to other ototoxic agents were analyzed.

**RESULTS:** Twenty-eight patients were included, with a mean age of 7.2 years at the beginning of chemotherapy (range 5 months-15 years 2 months); twenty-one patients received cisplatin, four received carboplatin, and three received both agents. Twelve patients had cranial irradiation and seven received another ototoxic medication. The most frequent malignancies were germ cell tumors, medulloblastoma and gliomas. Sensorineural hearing loss occurred in 28.6% of the patients with a mean follow-up period of 21.5 months (range: 1-53 months). All patients evaluated with audiometry had  $\geq$  Chang 2b ototoxicity. Risk factors include age less than 5 years, cranial irradiation, and cisplatin cumulative dose greater than 400 mg/m<sup>2</sup>.

**CONCLUSION:** Sensorineural hearing loss is a potential side effect of platinum-based chemotherapy. Pediatric patients receiving cisplatin chemotherapy with a cumulative dose exceeding 400 mg/m<sup>2</sup>, cranial irradiation as well as patients younger than 5 years are at greater risk of developing hearing loss.

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

[Prolonged furosemide exposure and risk of abnormal newborn hearing screen in premature infants. Early Hum Dev.](#)

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

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

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

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

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

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

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

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

**Wang X1, Huang L1, Zhao X1, Wang X1, Cheng X1, Du Y, Liu D.**

[Children with GJB2 gene mutations have various audiological phenotypes. Biosci Trends.](#)

2018 Sep 19;12(4):419-425. doi: 10.5582/bst.2018.01159. Epub 2018 Aug 27.

The current study retrospectively investigated variations in audiological phenotypes in children with GJB2 gene mutations. Subjects were 128 infants and young children who were seen as outpatients by Otolaryngology at Beijing Tongren Hospital from 2012 to 2018. Of the 128 subjects, 99 had biallelic truncating (T/T) mutations and 29 had truncating/nontruncating (T/NT) mutations. Genotypes, results of universal newborn hearing screening (UNHS), and the degree and symmetry of hearing loss were examined in the two groups. Twenty-two subjects (20.37%, 22/128) passed UNHS, including 13 children with T/T mutations and 9 with T/NT mutations. Of the 128 subjects, 22 had normal hearing, 2 had unilateral hearing loss, and 115 had bilateral hearing loss. Severe-to-profound hearing loss was the most prevalent phenotype in children with T/T mutations (73.23%), while normal hearing was prevalent in children with T/NT mutations (41.38%). Symmetrical hearing loss was the main phenotype in both groups, and the number of subjects with symmetrical hearing loss did not differ significantly between the two groups. Therefore, children with GJB2 gene mutations have phenotypic variability in terms of their results of UNHS and their degree and symmetry of hearing loss. Subjects with T/NT mutations of the GJB2 gene were more likely to pass UNHS and had milder hearing loss compared to those with T/T mutations. Symmetrical hearing loss was the main phenotype in the two groups, but 36.53% of children had bilateral asymmetric hearing loss. Parents of all subjects with sensorineural hearing loss were informed that their children may have a GJB2 mutation.

**Weber BC, Whitlock SM, He K, Kimbrell BS, Derkay CS.**

[An evidence based protocol for managing neonatal middle ear effusions in babies who fail newborn hearing screening. Am J Otolaryngol.](#)

2018 Sep - Oct;39(5):609-612. doi: 10.1016/j.amjoto.2018.04.004. Epub 2018 Apr 12.

**OBJECTIVES:** To evaluate the prevalence of middle ear disease in infants referred for failed newborn hearing screening (NBHS) and to review patient outcomes after intervention in order to propose an evidence-based protocol for management of newborns with otitis media with effusion (OME) who fail NBHS.

**METHODS:** 85 infants with suspected middle ear pathology were retrospectively reviewed after referral for failed NBHS. All subjects underwent a diagnostic microscopic exam with myringotomy with or without placement of a ventilation tube in the presence of a middle ear effusion and had intra-operative auditory brainstem response (ABR) testing or testing at a later date.

**RESULTS:** At the initial office visit, a normal middle ear space bilaterally was documented in 5 babies (6%), 29/85 (34%) had an equivocal exam while 51/85 (60%) had at least a unilateral OME. Myringotomy with or without tube placement due to presence of an effusion was performed on 65/85 (76%) neonates. Normal hearing was established in 17/85 (20%) after intervention, avoiding the need for any further audiologic workup. Bilateral or unilateral sensorineural hearing loss (SNHL) or mixed hearing loss was noted in 54/85 (64%) and these children were referred for amplification. Initially observation with follow up outpatient visits was initiated in 27/85 (32%) however, only 3/27 (11%) resolved with watchful waiting and 24/27 (89%) ultimately required at least unilateral tube placement due to OME and 14/24 (59%) were found to have at least a unilateral mixed or SNHL.

**CONCLUSIONS:** An effective initial management plan for children with suspected middle ear pathology and failed NBHS is diagnostic operative microscopy with placement of a ventilation tube in the presence of a MEE along with either intra-operative ABR or close follow-up ABR. This allows for the identification and treatment of babies with a conductive component due to OME, accurate diagnosing of an underlying SNHL component and for prompt aural rehabilitation.

**Wentland CJ, Ronner EA, Basonbul RA, Pinnapureddy S, Mankarious L, Keamy D, Lee DJ, Cohen MS.**

[Utilization of diagnostic testing for pediatric sensorineural hearing loss. Int J Pediatr Otorhinolaryngol.](#)

2018 Aug;111:26-31. doi: 10.1016/j.ijporl.2018.05.024. Epub 2018 May 22.

**BACKGROUND/OBJECTIVE:** Sensorineural hearing loss is a common diagnosis among children. The diagnostic workup varies widely among practitioners. This study's aim was to assess the utilization of diagnostic testing for SNHL and determine the yield of each test.

**STUDY DESIGN:** Retrospective chart review.

**SETTING:** Tertiary care center.

**SUBJECTS:** 827 patients with a diagnosis of SNHL from January 1, 2011 to January 1, 2015.

**RESULTS:** 746 patients met inclusion criteria. Temporal bone imaging was performed on 561 (75%) of patients with 224 (40%) having positive results that explained the etiology of the SNHL. Congenital SNHL was more likely to be associated with abnormal imaging than acquired SNHL (109/299 versus 106/316 respectively) ( $p = 0.001$ ). Unilateral SNHL was more likely to be associated with imaging abnormalities than bilateral SNHL (101/221 and 123/340 respectively) ( $p = 0.028$ ). Genetic testing was performed on 244 (33%) patients, of which 94 (39%) had abnormalities. Positive genetics results were more common with bilateral than unilateral SNHL (82/191 and 12/53 respectively) ( $p = 0.007$ ). There was no statistically significant difference in

the utility of genetic testing for congenital and acquired SNHL ( $p = 0.0836$ ). Cytomegalovirus (CMV) testing was available for 104 (14%) of patients with 13 (12.5%) being positive and consistent with congenital CMV. Electrocardiogram, urinalysis, and Lyme titers were less useful.

**CONCLUSIONS:** Imaging and genetic testing had the highest yield in the evaluation of children with SNHL and were the most commonly performed. CMV testing was valuable in neonates who failed newborn hearing screening.

Zahed Pasha Y, Zamani M, Hashemi Fard A, Zahed Pasha E.

[Screening of Hearing in Newborn Infants: Follow-Up and Outcome After 40930 Births in Babol, Northern Iran. Arch Iran Med.](#)

2018 Sep 1;21(9):382-386.

**BACKGROUND:** The purpose of this study was to investigate the results of hearing screening in all newborn infants, and their follow-up in Babol, northern Iran.

**METHODS:** Between 2006 and 2014, all healthy neonates delivered in 3 hospitals were included in this cross-sectional study. Newborns were screened using the transient evoked otoacoustic emissions test before discharge. Those who failed to pass the examination were tested for auditory brainstem response (ABR) by the age of one month. The infants referred from the previous level underwent tests of auditory steady state response, ABR and impedance audiometry before the age of 3 months. For infants with the diagnosis of bilateral hearing impairment, it was recommended to use a hearing aid in 3 months. Then, their parents were recommended to take infants again to the hearing testing centers within next 6 months. If the infant's hearing was not improved, he/she was advised to undergo cochlear implantation.

**RESULTS:** In total, 40930 newborns were screened. Out of them, 62 (1.5 per 1000 live births) were finally diagnosed to have hearing impairment, of whom 14 had unilateral and 48 had bilateral disorders (candidate for supportive measures). Overall, 986 (2.4%) were lost to follow-up and 11 (0.03%) died over the first 3 months of age. At the end of the 6-month follow-up for supportive stage, 15 out of 48 infants received a hearing aid and 18 (0.4 per 1000 children) underwent cochlear implant surgery. Fourteen out of 48 cases were lost to follow-up over supportive stage.

**CONCLUSION:** It is recommended that all newborns undergo hearing screening test before hospital discharge, and those with impairment receive supportive measures from 3 months of age, and be re-examined at 12 months of age.

Zhao P, Lin L, Lan L.

[Analysis of mutation spectrum of common deafness-causing genes in Hakka newborns in southern China by semiconductor sequencing. Medicine \(Baltimore\).](#)

2018 Sep;97(38):e12285. doi: 10.1097/MD.00000000000012285.

Hearing loss is a common neurosensory disorder, approximately half of the cases are caused by genetic factors, and approximately 70% of hereditary hearing impairments are nonsyndromic hearing loss (NSHL). The mutations of GJB2 (gap junction beta-2 protein), GJB3 (gap junction beta-3 protein), SLC26A4 (solute carrier family 26 member 4), and MT-RNR1 (mitochondrially encoded 12S RNA) are the most common inherited causes of NSHL. Because of different genetic backgrounds, the mutation spectrum of these common deafness-causing genes varies among different regions in China. Because no data are known on these mutations among the Hakka population of Southern China, we aim to investigate the mutation spectrum to add these to neonatal screening and genetic counseling. A total of 1252 blood samples from newborns have been detected by semiconductor sequencing for 100 mutations loci of 18 deafness-causing genes. Of the participants, 95 subjects carried deafness-causing genes mutations with the carrier rate of 7.59%. The mutation frequencies of GJB2, SLC26A4, GJB3, and mitochondrial genes were 3.04%, 3.51%, 0.16%, and 0.88%, respectively. We followed up subjects with single-gene homozygous or compound heterozygous mutations. Our study firstly analyzed deafness-causing genes mutation spectrum in Hakka population, providing evidence for future neonatal screening and genetic counseling in this area.

Zhao XL, Huang LH, Wang XY, DU Y, Wang X, Cheng XH, Zhao LP, Li Y.

[Analysis of genotypes and audiological characteristics of children with SLC26A4 gene pathogenic mutations. Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi.](#)

2018 Jun 5;32(11):836-840. doi: 10.13201/j.issn.1001-1781.2018.11.009.[Article in Chinese]

**Objective:**To explore the correlation of SLC26A4 genotype and audiology.**Method:**The subjects were 70 children aged 0 to 7 years old, who were admitted to otological outpatient department.All subjects received nine crystal hereditary deafness gene chip and confirmed by (or)SLC26A4 gene full coding region detection.The patients were diagnosed as homozygous or compound heterozygous mutations.At the same time,acoustic immittance,auditory brainstem response, auditory steady state response and pediatric behavior audiometry, newborn hearing screening and other audiological tests were displayed. According to the genotype, the subjects were divided into two groups: group A (SLC26A4 gene homozygous mutation) in 40 cases, group B (SLC26A4 gene compound heterozygous mutation) in 30 cases. The frequency of SLC26A4 gene mutation, the two groups of genotypes and hearing screening results,the degree of hearing loss and audiometric configurations were analyzed statistically. **Result:** In 70 patients, the top 4 of the 70 patients with high frequency of mutations were IVS7-2A> G(76.43%), 2168A> G(15.00%), 1226G> A(2.86%) and 2000T> C(2.16%), respectively. 34.29% of newborns passed hearing screening with single or double ears, among which group A and group B were 32.50% and 36.67%,respectively. There was no statistically significant difference between two groups in hearing screening. The degree of hearing loss in group A(56.25%) and group B(48.33%) were mainly profound and there was no significant difference between them. The audiometric configurations: group A(60.00%) was mainly high frequency loss type, while group B(55.00%) was mainly flat type. The difference between them was statistically significant.**Conclusion:**The mutation sites of SLC26A4 gene were mainly IVS7-2A> G, and the degree of hearing loss was mostly profound. To the audiometric configurations,SLC26A4 gene homozygous mutant were mainly high frequency loss type, while SLC26A4 gene compound heterozygous mutant were mainly flat type. 34.29% children passed universal newborn hearing screening with one ear at least, which indicates SLC26A4 gene mutations can result in late-onset hearing loss, so those patients should be attached great importance.