

Others' Publications About EHDI: May through October 2017

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.

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

It is interesting to note that of the 80 articles published in other journals from May through October 2017, 59 articles (74%) reported on research conducted in countries other than the United States. Clearly, EHDI has become a global phenomenon and keeping up with EHDI requires staying informed about what is happening in other countries. As an indicator of what is trending in the literature:

- 23 articles discuss issues related to cochlear implants.
- 11 articles explore issues related to the genetics of hearing loss.
- 8 articles discuss the identification or treatment of cytomegalovirus (CMV) and its relation to childhood hearing loss.
- 7 articles report on issues related to protocols and procedures used in newborn hearing screening.

Noted below are just some of the interesting findings from around the world.

- Chung et. al surveyed 1024 audiology facilities in the United States and found that more than 90% reported results of diagnostic evaluations following newborn hearing screening to their state EHDI program.
- Fitzpatrick et. al reported that 16.5% of children identified with hearing loss in their sample from Canada experienced more than 3 months delay from the initial audiologic assessment to confirmation of their hearing disorder.
- Martinez-Cruz et. al found that 40% of the Mexican infants from neonatal intensive care units with sensorineural hearing loss had progressive hearing loss.
- Netten et. al reported that children from the Netherlands and Belgium who were identified early and received cochlear implants had higher early language skills, which served as a protective factor against the development of disruptive behavior.
- Sanyelbhaa et. al found a 76% increased risk for sensorineural hearing loss among children in Egypt who were progeny of consanguineous marriages compared to similar children from non-consanguineous marriages.
- Guerzoni and Cuda used data logging from cochlear implant speech processors to show that the amount of time the cochlear implant was worn positively predicted early linguistic and auditory outcomes.

Listed below are many more articles with interesting and relevant findings that can be used to improve EHDI programs wherever you live.

Abdul Majid AH, Zakaria MN, Abdullah NAC, Hamzah S, Mukari SZS.

[Determinants of caregivers' awareness of Universal Newborn Hearing Screening in Malaysia.](#)

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:107-111. doi: 10.1016/j.ijporl.2017.07.036. Epub 2017 Jul 27.

OBJECTIVE: This paper aims to investigate the effects of perceived attitude and anxiety on awareness of UNHS among caregivers in Malaysia.

METHODS: Using cross sectional research approach, data were collected and some 46 out of 87 questionnaires distributed to caregivers attending UNHS programs at selected public hospitals were usable for analysis (response rate of 52.8%). Partial Least Squares Method (PLS) algorithm and bootstrapping technique were employed to test the hypotheses of the study.

RESULTS: R square value is 0.205, and it implies that exogenous latent variables explained 21% of the variance of the endogenous latent variable. This value indicates moderate and acceptable level of R-squared values. Findings from PLS structural model evaluation revealed that anxiety has no significant influence ($\beta = -0.091$, $t = 0.753$, $p > 0.10$) on caregivers' awareness; but perceived attitude has significant effect ($\beta = -0.444$, $t = 3.434$, $p < 0.01$) on caregivers' awareness.

CONCLUSION: Caregivers' awareness of UNHS is influenced by their perceived attitude while anxiety is not associated with caregivers' awareness. This implies that caregivers may not believe in early detection of hearing impairment in children, thinking that their babies are too young to be tested

for hearing loss. Moreover, socio-economic situation of the caregivers may have contributed to their failure to honor UNHS screening appointments as some of them may need to work to earn a living while some may perceive it a waste of time honoring such appointments. Non-significant relationship between anxiety and caregivers' awareness may be due to religious beliefs of caregivers. Limitations and suggestions were discussed.

Aimoni C, Ciorba A, Cerritelli L, Ceruti S, Skarzyński PH, Hatzopoulos S.

Enlarged vestibular aqueduct: Audiological and genetical features in children and adolescents.

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:254-258. doi: 10.1016/j.ijporl.2017.07.042. Epub 2017 Jul 29.

BACKGROUND: Enlarged Vestibular Aqueduct (EVA) is one of the most common congenital malformations associated with sensorineural or mixed hearing loss. The association between hearing loss and EVA is described in syndromic (i.e. Pendred Syndrome, BOR, Waardenburg) and non-syndromic disorders, as isolate or familiar mutations of the SLC26A4 gene. The audiological phenotype of the EVA syndrome is heterogeneous, the type and entity of hearing loss may vary and vertigo episodes might also be present.

OBJECTIVE: The aim of this retrospective study was to describe the clinical and genetic features of a group of adolescent subjects presenting an EVA clinical profile, considering the presence of SLC26A4 gene mutations.

METHODS: 14 Caucasian patients were assessed (24 ears in total; 4 patients presented a monolateral EVA), 10 females and 4 males. Their age at the time of diagnosis was between 1 and 6 years (mean age 2.5 years). Subjects were assessed by an ENT microscopy evaluation with a complete audiometric assessment, CT & MRI scans and genetic tests for the evaluation of the pendrin gene mutations (SLC26A4).

RESULTS: Considering the presence of SLC26A4 mutations and thyroid function, we could identify three sub-groups of patients: group 1, non syndromic EVA (ns EVA, no SLC26A4 mutation and no thyroid dysfunction); group 2, EVA with DFNB4 (single SLC26A4 gene mutation and no thyroid dysfunction); group 3, EVA with Pendred Syndrome (two pathological mutation of SLC26A4 and thyromegaly with thyroid dysfunction). Patients of group 1 (ns-EVA) showed various degrees of hearing loss from mild (55%) to severe-profound (45%). In groups 2 (DFNB4) and 3 (PDS), the degree of hearing loss is severe to profound in 70-75% of the cases; middle and high frequencies are mainly involved.

CONCLUSIONS: The phenotypic expressions associated with the EVA clinical profile are heterogeneous. From the available data, it was not possible to identify a representative audiological profile, in any of the three sub-groups. The data suggest that: (i) a later onset of hearing loss is usually related to EVA, in absence of SLC26A4 gene mutations; and (ii) hearing loss is more severe in patients with SLC26A4 gene mutations (groups 2 and 3 of this study).

Al-Sayed AA, AlSanosi A.

Cochlear implants in children: A cross-sectional investigation on the influence of geographic location in Saudi Arabia

J Family Community Med. 2017 May-Aug;24(2):118-121. doi: 10.4103/jfcm.JFCM_142_15.

INTRODUCTION: The role of the family in detecting a child's hearing difficulty and the age at which an implantation is done have been identified as strong predictors of the outcomes of pediatric cochlear implantation. In the absence of screening programs for hearing loss in Saudi neonates, the family's role is of paramount importance. The aim of this study was to investigate the influence of geographic location on the course of identification, examination, and cochlear implantation in children in Saudi Arabia.

MATERIALS AND METHODS: Pediatric patients who had received either unilateral or bilateral cochlear implantation at King Abdulaziz University Hospital in Riyadh, Saudi Arabia, between January 1, 2012, and December 31, 2014, were surveyed.

RESULTS: A total of 156 pediatric patients have had a cochlear implant between January 1, 2012, and December 31, 2014. The one-way analysis of variance test to compare the means of the independent sample groups in various geographic zones showed that with a hundred percent access to primary health care, the geographic location of the population had an influence on the detection of hearing loss but not on the cochlear implantation.

CONCLUSION: This study found that the geographic location of the population has an influence on the time of detection of hearing loss in children but not on the time of cochlear implantation. Raising parental awareness of the importance of early detection of hearing loss is necessary. Further research is also required to define the role of factors such as the income and the educational level of parents on the early detection of neonatal hearing loss.

Alkowari MK, Vozzi D, Bhagat S, Krishnamoorthy N, Morgan A, Hayder Y, Logendra B, Najjar N, Gandin I, Gasparini P, Badii R, Giroto G, Abdulhadi K.

Targeted sequencing identifies novel variants involved in autosomal recessive hereditary hearing loss in Qatari families.

Mutat Res. 2017 Aug;800-802:29-36. doi: 10.1016/j.mrfmmm.2017.05.001. Epub 2017 May 4.

Hereditary hearing loss is characterized by a very high genetic heterogeneity. In the Qatari population the role of GJB2, the worldwide HHL major player, seems to be quite limited compared to Caucasian populations. In this study we analysed 18 Qatari families affected by non-syndromic hearing loss using a targeted sequencing approach that allowed us to analyse 81 genes simultaneously. Thanks to this approach, 50% of these families (9 out of 18) resulted positive for the presence of likely causative alleles in 6 different genes: CDH23, MYO6, GJB6, OTOF, TMC1 and OTOA. In particular, 4 novel alleles were detected while the remaining ones were already described to be associated to HHL in other ethnic groups. Molecular modelling has been used to further investigate the role of novel alleles identified in CDH23 and TMC1 genes demonstrating their crucial role in Ca²⁺ binding and therefore possible functional role in proteins. Present study showed that an accurate molecular diagnosis based on next generation sequencing technologies might largely improve molecular diagnostics outcome leading to benefits for both genetic counseling and definition of recurrence risk.

Amin SB, Saluja S, Saili A, Orlando M, Wang H, Laroia N, Agarwal A.

Chronic auditory toxicity in late preterm and term infants with significant hyperbilirubinemia.

Pediatrics. 2017 Oct;140(4). pii: e20164009. doi: 10.1542/peds.2016-4009.

BACKGROUND AND OBJECTIVES: Significant hyperbilirubinemia (SHB) may cause chronic auditory toxicity (auditory neuropathy spectrum disorder and/or sensorineural hearing loss); however, total serum bilirubin (TSB) does not discriminate neonates at risk for auditory toxicity. Our objective was to compare TSB, bilirubin albumin molar ratio (BAMR), and unbound bilirubin (UB) for their association with chronic auditory toxicity in neonates with SHB (TSB \geq 20 mg/dL or TSB that met criteria for exchange transfusion).

METHODS: Infants \geq 34 weeks' gestational age (GA) with SHB during the first 2 postnatal weeks were eligible for a prospective longitudinal study in India. Comprehensive auditory evaluations were performed at 2 to 3 months of age by using auditory brainstem response, tympanometry, and an otoacoustic emission test and at 9 to 12 months of age by using audiometry. The evaluations were performed by an audiologist unaware of the degree of jaundice.

RESULTS: A total of 93 out of 100 infants (mean GA of 37.4 weeks; 55 boys, 38 girls) who were enrolled with SHB were evaluated for auditory toxicity. Of those, 12 infants (13%) had auditory toxicity. On regression analysis controlling for covariates, peak UB (but not peak TSB or peak BAMR), was

associated with auditory toxicity (odds ratio 2.41; 95% confidence interval: 1.43-4.07; $P = .001$). There was significant difference in the area under the receiver operating characteristic curves between UB (0.866), TSB (0.775), and BAMR (0.724) for auditory toxicity ($P = .03$) after controlling for covariates.

CONCLUSIONS: Unconjugated hyperbilirubinemia indexed by UB (but not TSB or BAMR) is associated with chronic auditory toxicity in infants ≥ 34 weeks' GA with SHB.

Ari-Even Roth D, Lubin D, Kuint J, Teperberg-Oikawa M, Mendelson E, Strauss T, Barkai G.

Contribution of targeted saliva screening for congenital CMV-related hearing loss in newborns who fail hearing screening.

Arch Dis Child Fetal Neonatal Ed. 2017 Nov;102(6):F519-F524. doi: 10.1136/archdischild-2016-311859. Epub 2017 May 3.

BACKGROUND: We previously reported a 2.2% rate of infants born with sensorineural hearing loss (SNHL) due to congenital cytomegalovirus (cCMV) infection identified by universal neonatal screen for cCMV using saliva.

OBJECTIVE: To evaluate the contribution of targeted saliva screening for cCMV to the detection of infants born with cCMV-related SNHL who failed universal newborn hearing screening (UNHS).

METHODS: We retrospectively reviewed the audiological and medical records of infants who failed UNHS and were tested for cCMV using saliva sample prior to discharge at Sheba Medical Center between 2014 and 2015. Positive cases were confirmed by urine sample.

RESULTS: Two hundred (1%) of the 19830 infants tested during the study period failed in-hospital hearing screening. A saliva specimen was obtained prior to discharge in 187 infants (93.5% of those who failed UNHS). In 178 infants saliva testing was performed at ≤ 21 days of chronological age and yielded results. cCMV infection was identified in 4/178 tested infants (2.25%, 95% CI 0.8% to 5.3%), of whom three were diagnosed with SNHL (1.7%, 95% CI 0.5% to 4.4%) and offered antiviral treatment. Two of the tested infants (1.12%, 95% CI 0.2% to 3.6%) were diagnosed with cCMV solely due to failure in UNHS. Occult central nervous system (CNS) symptoms of cCMV infection were detected in 2/4 infants following targeted investigation.

CONCLUSIONS: Targeted cCMV screening in newborns who failed UNHS contributed to the early detection of infants born with cCMV-related isolated SNHL or with occult CNS symptoms who could potentially benefit from antiviral treatment.

Bernardi GF, Pires CTF, Oliveira NP, Nisihara R.

Prevalence of pressure equalization tube placement and hearing loss in children with Down Syndrome.

Int J Pediatr Otorhinolaryngol. 2017 Jul;98:48-52. doi: 10.1016/j.ijporl.2017.04.041. Epub 2017 Apr 27.

OBJECTIVE: To determine the prevalence of pressure equalization tube (PET) placement and hearing loss in children with Down syndrome (DS).

MATERIAL AND METHODS: We evaluated 90 DS children births between 1 and 11 years old and compared to 90 children without DS paired in sex and age. Medical records were analyzed consecutively. We collected data about proceedings PET placement, age of the patient at each PET, adenoidectomy, tonsillectomy and results for audiometry and tympanometry.

RESULTS: Among the 90 patients with DS, 49 (54.4%) were male, median age of 58 months (15-143 months). In this group, 75 PET were placed in 26/90 children (28.9%) mostly between 3 and 5 years old. In 10/26 (38.5%) was necessary PET replaced. When compared to the control group- 6/90 (6.7%)- children with DS presented OR = 13.7 (95% CI 4.0-47.3) times more likely to use PET. Adenoidectomy and tonsillectomy (44.4% and 42.2% respectively) were significantly more frequent in DS group. The prevalence of hearing loss was 32.1% in the right ear and 26.9% in the left ear. Type B tympanometry was found in more than half of the patients with DS.

CONCLUSION: We found a 13-fold higher risk of PET in DS children, especially between the ages of 3-5 years. The high prevalence of hearing loss and PET placement in patients with DS reinforcing the importance of early and regular follow-up for hearing screening in this population, mostly in preschool-aged children.

Berrettini S, Ghirri P, Lazzarini F, Lenzi G, Forli F.

Newborn hearing screening protocol in Tuscany region.

Ital J Pediatr. 2017 Sep 20;43(1):82. doi: 10.1186/s13052-017-0397-1.

BACKGROUND: Newborn hearing screening has to be considered the first step of a program for the identification, diagnosis, treatment and habilitation/rehabilitation of children with hearing impairment.

MAIN PART: In Tuscany Region of Italy, the universal newborn hearing screening is mandatory since November 2007. The first guidelines for the execution of the screening have been released in June 2008; then many other Italian regions partially or totally adopted these guidelines. On the basis of the experience from 2008 and according to the recent evidences in the scientific literature, a new screening protocol was released in Tuscany region. The new protocol is an evolution of the previous one. Some issues reported in the previous protocol and in the Joint Committee on Infant Hearing statement published in 2007 were revised, such as the risk factors for auditory neuropathy and for late onset, progressive or acquired hearing loss. The new updated guidelines were submitted to the Sanitary Regional Council and then they have been approved in August 2016. The updated screening protocol is mainly aimed to identify newborns with a congenital moderate-to-profound hearing loss, but it also provides indications for the audiological follow-up of children with risk's factor for progressive or late onset hearing loss; further it provides indications for the audiological surveillance of children at risk for acquired hearing impairment. Then, in the new guidelines the role of the family paediatrician in the newborn hearing screening and audiological follow-up and surveillance is underscored. Finally the new guidelines provide indications for the treatment with hearing aids and cochlear implant, in accordance with the recent Italian Health Technology Assessment (HTA) guidelines.

CONCLUSIONS: In the paper we report the modality of execution of the universal newborn hearing screening in the Tuscany Region, according to the recently updated protocol. The main features of the protocol and the critical issues are discussed.

Blázquez-Gamero D, Galindo Izquierdo A, Del Rosal T, Baquero-Artigao F, Izquierdo Méndez N, Soriano-Ramos M, Rojo Conejo P, González-Tomé MI, García-Burguillo A, Pérez Pérez N, Sánchez V, Ramos-Amador JT, De la Calle M.

Prevention and treatment of fetal cytomegalovirus infection with cytomegalovirus hyperimmune globulin: a multicenter study in Madrid.

J Matern Fetal Neonatal Med. 2017 Oct 4:1-211. doi: 10.1080/14767058.2017.1387890. [Epub ahead of print]

INTRODUCTION: Cytomegalovirus (CMV) is the leading cause of congenital infection worldwide. Data about the management of CMV infection in pregnant women are scarce, and treatment options are very limited. The aim of the study is to investigate the effectiveness of cytomegalovirus hyperimmune globulin (CMV-HIG) for the prevention and treatment of congenital CMV (cCMV) infection.

MATERIALS AND METHODS: A retrospective observational study was conducted in three tertiary hospitals in Madrid. In the period 2009-2015, CMV-HIG (Cytotect® CP Biotest, Biotest) treatment was offered to all pregnant women with primary CMV infection and/or detection of CMV-DNA in amniotic fluid in participating centers. Women were divided into prevention and treatment groups (PG and TG, respectively). Those with primary CMV infection who had not undergone amniocentesis comprised the PG and received monthly CMV-HIG (100 UI/kg). If CMV-DNA was subsequently detected in

amniotic fluid, one extra dose of CMV-HIG (200 UI/kg) was given 4 weeks after the last dose. Those women were considered to be part of the PG group despite detection of CMV-DNA in amniotic fluid. In the case of a negative result in CMV-DNA detection in amniotic fluid or if amniocentesis was not performed, monthly HIG was given up to the end of the pregnancy.

RESULTS: Thirty-six pregnant women were included. Median gestational age at birth was 39 weeks [interquartile range (IQR): 38-40] and 2 children (5.5%) were premature (born at 28 and 34 weeks' gestation). Amniocentesis was performed in 30/36 (83.4%) pregnancies and CMV PCR was positive in 21 of them (70%). One fetus with a positive PCR in amniotic fluid that received one dose of HIG after amniocentesis presented a negative CMV-PCR in urine at birth, and was asymptomatic at 12 months of age. Twenty-four children were infected at birth, and 16/21 (76.2%) presented no sequelae at 12 months, while 2 (9.5%) had mild unilateral hearing loss and three (14.3%) severe hearing loss or neurological sequelae. Seventeen women were included in the PG and 19 in the TG. In the PG 7/17 (41%) fetuses were infected, one pregnancy was terminated due to abnormalities in cordocentesis and one showed mild hearing loss at 12 months of age. In the TG, 18/19 children (95%) were diagnosed with cCMV, while the remaining neonate had negative urine CMV at birth. Eight out of the 19 fetuses (42.1%) showed CMV related abnormalities in fetal US before HIG treatment. Complete clinical assessment in the neonatal period and at 12 months of age was available in 16 and 15 children, respectively. At birth 50% were symptomatic and at 12 months of age, 4/15 (26.7%) showed hearing loss and 3/15 (20%) neurologic impairment. Fetuses with abnormalities in ultrasonography before HIG presented a high risk of sequelae (odds ratios (OR): 60 95%CI: 3-1185; $p = 0.007$).

DISCUSSION: Prophylactic HIG administration in pregnant women after CMV primary infection seems not to reduce significantly the rate of congenital infection, but is safe and it could have a favorable effect on the symptoms and sequelae of infected fetuses. The risk of long-term sequelae in fetuses without US abnormalities before HIG is low, so it could be an option in infected fetuses with normal imaging. On the other hand, the risk of sequelae among infected fetuses with abnormalities in fetal ultrasonography (FUS) before HIG despite treatment is high.

Bruijnzeel H, Bezdjian A, Lesinski-Schiedat A, Illg A, Tzifa

[Evaluation of pediatric cochlear implant care throughout Europe: Is European pediatric cochlear implant care performed according to guidelines?](#)

Cochlear Implants Int. 2017 Nov;18(6):287-296. doi: 10.1080/14670100.2017.1375238. Epub 2017 Sep 19.

OBJECTIVES: International guidelines indicate that children with profound hearing loss should receive a cochlear implant (CI) soon after diagnosis in order to optimize speech and language rehabilitation. Although prompt rehabilitation is encouraged by current guidelines, delays in cochlear implantation are still present. This study investigated whether European countries establish timely pediatric CI care based on epidemiological, commercial, and clinical data.

METHODS: An estimation of the number of pediatric CI candidates in European countries was performed and compared to epidemiological (Euro-CIU), commercial (Cochlear®), and clinical (institutional) age-at-implantation data. The ages at implantation of pediatric patients in eight countries (the Netherlands, Belgium, Germany, the United Kingdom, France, Turkey, Portugal, and Italy) between 2005 and 2015 were evaluated.

RESULTS: From 2010 onwards, over 30% of the pediatric CI candidates were implanted before 24 months of age. Northern European institutions implanted children on average around 12 months of age, whereas southern European institutions implanted children after 18 months of age. The Netherlands and Germany implanted earliest (between 6 and 11 months).

DISCUSSION: Implemented newborn hearing screening programs and reimbursement rates of CIs vary greatly within Europe due to local, social, financial, and political differences. However, internationally accepted recommendations are applicable to this heterogeneous European CI practice. Although consensus on early pediatric cochlear implantation exists, this study identified marked delays in European care.

CONCLUSION: Regardless of the great heterogeneity in European practice, reasons for latency should be identified on a national level and possibilities to prevent avoidable future implantation delays should be explored to provide national recommendations.

Bush ML, Taylor ZR, Noblitt B, Shackelford T, Gal TJ, Shinn JB, Creel LM, Lester C, Westgate PM, Jacobs JA, Studts CR.

[Promotion of early pediatric hearing detection through patient navigation: A randomized controlled clinical trial.](#)

Laryngoscope 2017 Nov;127 Suppl 7:S1-S13. doi: 10.1002/lary.26822. Epub 2017 Sep 20.

OBJECTIVES/HYPOTHESIS: To assess the efficacy of a patient navigator intervention to decrease nonadherence to obtain audiological testing following failed screening, compared to those receiving the standard of care.

METHODS: Using a randomized controlled design, guardian-infant dyads, in which the infants had abnormal newborn hearing screening, were recruited within the first week after birth. All participants were referred for definitive audiological diagnostic testing. Dyads were randomized into a patient navigator study arm or standard of care arm. The primary outcome was the percentage of patients with follow-up nonadherence to obtain diagnostic testing. Secondary outcomes were parental knowledge of infant hearing testing recommendations and barriers in obtaining follow-up testing.

RESULTS: Sixty-one dyads were enrolled in the study (patient navigator arm=27, standard of care arm=34). The percentage of participants nonadherent to diagnostic follow-up during the first 6 months after birth was significantly lower in the patient navigator arm compared with the standard of care arm (7.4% vs. 38.2%) ($P = .005$). The timing of initial follow-up was significantly lower in the navigator arm compared with the standard of care arm (67.9 days after birth vs. 105.9 days, $P = .010$). Patient navigation increased baseline knowledge regarding infant hearing loss diagnosis recommendations compared with the standard of care ($P = .004$).

CONCLUSIONS: Patient navigation decreases nonadherence rates following abnormal infant hearing screening and improves knowledge of follow-up recommendations. This intervention has the potential to improve the timeliness of delivery of infant hearing healthcare; future research is needed to assess the cost and feasibility of larger scale implementation.

Carew P, Mensah FK, Rance G, Flynn T, Poulakis Z, Wake M.

[Mild-moderate congenital hearing loss: Secular trends in outcomes across four systems of detection.](#)

Child Care Health Dev. 2017 Jun 14. doi: 10.1111/cch.12477. [Epub ahead of print]

BACKGROUND: Universal newborn hearing screening (UNHS) targets moderate or greater hearing loss. However, UNHS also frequently detects children with mild loss that results in many receiving early treatment. The benefits of this approach are not yet established. We aimed to (i) compare language and psychosocial outcomes between four hearing loss detection systems for children aged 5-8 years with congenital mild-moderate hearing loss; (ii) determine whether age of detection predicts outcomes; and (iii) compare outcomes between children identified via well-established UNHS and the general population.

METHODS: Linear regression adjusted for potential confounding factors was used throughout. Via a quasi-experimental design, language and psychosocial outcomes were compared across four population-based Australian systems of hearing loss detection: opportunistic detection, born 1991-1993, $n = 50$; universal risk factor referral, born 2003-2005, $n = 34$; newly established UNHS, born 2003-2005, $n = 41$; and well-established UNHS, born 2007-2010, $n = 21$. In pooled analyses, we examined whether age of detection predicted outcomes. Outcomes were similarly compared between

the current well-established UNHS system and typically developing children in the Early Language in Victoria Study, born 2003, n = 1217.

RESULTS: Age at diagnosis and hearing aid fitting fell steadily across the four systems. For moderate losses, mean expressive language (P for trend .05) and receptive vocabulary (P for trend .06) improved across the four systems, but benefit was not obvious for mild losses. In pooled analyses, diagnosis before age six months predicted better language outcomes for moderate losses. Children with mild-moderate losses exposed to well-established UNHS continue to experience expressive language scores well below children in the general population (adjusted mean difference -8.9 points, 95% CI -14.7 to -3.1).

CONCLUSIONS: Treatment arising from UNHS appears to be clearly benefitting children with moderate hearing losses. However, rigorous trials are needed to quantify benefits, versus costs and potential harms, of early aiding of children with mild losses.

Chan KH, Gao D, Jensen EL, Allen GC, Cass SP.

[Complications and parent satisfaction in pediatric osseointegrated bone-conduction hearing implants.](#)

Laryngoscope. 2017 Sep;127(9):2165-2170. doi: 10.1002/lary.26469. Epub 2017 Jan16.

OBJECTIVE: To assess long-term complication rate and parental satisfaction of osseointegrated bone conduction hearing implants (OBCHIs).

STUDY DESIGN: Retrospective chart review of children undergoing OBCHIs.

METHODS: A retrospective chart review of children undergoing OBCHIs for the treatment of conductive, mixed, and single-sided sensorineural hearing loss in children.

RESULTS: Forty-five subjects were identified with 0.3 to 10.4 years of follow-up. The mean/median age and age range at implant were 9.0/7.8 and 1.7 to 19.1 years. The underlying hearing loss for the cohort included conductive (N = 30), sensorineural (N = 7), and mixed (N = 8) hearing loss.

Conductive hearing loss, caused by aural atresia (62.9), was the most common indication for implantation. Fifty-eight complications occurred in 29 subjects, most related to skin infection or overgrowth. Seventeen events required revision surgery, and 18 required oral antibiotics and/or office-based cauterization. Children under the age of 5 years were more likely to have failure of osseointegration or require revision surgery. Parents of 33 subjects underwent a phone interview; 76% rated the overall satisfaction as satisfied or very satisfied.

CONCLUSION: A large percentage of children undergoing OBCHI develop postoperative complications, and up to 44% require revision surgery—a figure higher than generally reported and higher than in adults. No factors were found to adequately explain the higher complication rates in children compared to adults. Despite the occurrence of complications, parents viewed this device as satisfactory from many perspectives.

Chen X, Yuan M, Lu J, Zhang Q, Sun M, Chang F.

[Assessment of universal newborn hearing screening and intervention in Shanghai, China.](#)

Int J Technol Assess Health Care. 2017 Jan;33(2):206-214. doi:10.1017/S0266462317000344. Epub 2017 Jun 6.

OBJECTIVES: The aim of this study was to evaluate the universal newborn hearing screening (UNHS) and intervention program in Shanghai, China.

METHODS: This study included the quantitative analyses of the UNHS-Shanghai database in 2002-12 and qualitative assessment of the program.

The Otoacoustic Emissions and the Automated Auditory Brainstem Evoked Responses tests were conducted in screening. The costs and benefits were calculated based on the number of participants in each stage. The short-term and long-term periods were defined as from birth to 15 years of age or to death (82-year-olds), respectively. Sensitivity analyses were conducted.

RESULTS: A total 1,574,380 newborns were included, representing 93.6 percent of all eligible babies in Shanghai during the study period. The prevalence of newborn hearing loss was 1.66%. The short-term/long-term program costs were ¥488.5 million (US\$75.52 million)/¥1.08 billion (US\$167.12 million), and the short-term/long-term program benefit was ¥980.1 million (US\$151.53 million)/¥8.13 billion (US\$1.26 billion). The program benefit was greater than its cost if the proportion of hearing-loss children enrolled in regular schools was no less than 41.4 percent of all hearing impaired children, as well as if the wage growth rate ranged from 3 percent to 8 percent. Qualitative results also suggested that stakeholders strongly supported this program.

CONCLUSIONS: The universal newborn hearing screening and intervention program in Shanghai is justified in terms of the resource input in the long run, although there is still room for further improvement with respect to educational rehabilitation and a better infrastructure system.

Ching TYC, Dillon H, Button L, Seeto M, Van Buynder P, Marnane V, Cupples L, Leigh G.

[Age at intervention for permanent hearing loss and 5-year language outcomes.](#)

Pediatrics. 2017 Sep;140(3). pii: e20164274. doi: 10.1542/peds.2016-4274. Epub 2017 Aug 3.

OBJECTIVES: Universal newborn hearing screening has been implemented to detect permanent childhood hearing loss (PCHL) early, with the ultimate goal of improving outcomes through early treatment. However, there is disagreement between studies on the size of this benefit and in some cases whether it is significantly different from 0. There have been no studies of sufficient size in which researchers have determined reliably whether the effect varies with degree of PCHL. We aimed to explore how intervention timing influences 5-year language in children with PCHL.

METHODS: Via a prospective study of 350 children, we used standard multiple regression analyses to investigate the effect of age at intervention or hearing screening on language outcomes after allowing for the effects of nonverbal IQ, degree of PCHL, sex, birth weight, maternal education, additional disabilities, and communication mode.

RESULTS: The benefit of early intervention for language development increased as hearing loss increased. Children whose amplification started at age 24 months had poorer language than those whose amplification started at 3 months. The difference was larger for 70-dB HL (-11.8 score points; 95% confidence interval [95% CI]: -18.7 to -4.8) than for 50-dB HL (-6.8; 95% CI: -10.8 to -2.8). Children who received cochlear implants at 24 months had poorer language than those implanted at 6 months (-21.4; 95% CI: -33.8 to -9.0). There was no significant effect of screening on outcomes.

CONCLUSIONS: Early intervention improves language outcomes, thereby lending support to streamlining clinical pathways to ensure early amplification and cochlear implantation after diagnosis.

Chiossi JSC, Hyppolito MA.

[Effects of residual hearing on cochlear implant outcomes in children: A systematic-review.](#)

Int J Pediatr Otorhinolaryngol. 2017 Sep;100:119-127. doi: 10.1016/j.ijporl.2017.06.036. Epub 2017 Jul 1.

OBJECTIVES: to investigate if preoperative residual hearing in prelingually deafened children can interfere on cochlear implant indication and outcomes.

METHODS: a systematic-review was conducted in five international databases up to November-2016, to locate articles that evaluated cochlear implantation in children with some degree of preoperative residual hearing. Outcomes were auditory, language and cognition performances after cochlear implant. The quality of the studies was assessed and classified according to the Oxford Levels of Evidence table - 2011. Risk of biases were also described.

RESULTS: From the 30 articles reviewed, two types of questions were identified: (a) what are the benefits of cochlear implantation in children with residual hearing? (b) is the preoperative residual hearing a predictor of cochlear implant outcome? Studies ranged from 04 to 188 subjects, evaluating populations between 1.8 and 10.3 years old. The definition of residual hearing varied between studies. The majority of articles (n = 22) evaluated speech perception as the outcome and 14 also assessed language and speech production.

CONCLUSION: There is evidence that cochlear implant is beneficial to children with residual hearing. Preoperative residual hearing seems to be valuable to predict speech perception outcomes after cochlear implantation, even though the mechanism of how it happens is not clear. More extensive researches must be conducted in order to make recommendations and to set prognosis for cochlear implants based on children preoperative residual hearing.

Chung W, Beauchaine KL, Grimes A, O'Hollearn T, Mason C, Ringwalt S.

Reporting newborn audiologic results to state EHDI programs.

Ear Hear. 2017 Sep/Oct;38(5):638-642. doi: 10.1097/AUD.0000000000000443.

OBJECTIVES: All US states and territories have an Early Hearing Detection and Intervention (EHDI) program to facilitate early hearing evaluation and intervention for infants who are deaf or hard of hearing. To ensure efficient coordination of care, the state EHDI programs rely heavily on audiologists' prompt reporting of a newborn's hearing status. Several states have regulations requiring mandatory reporting of a newborn's hearing status. This is an important public health responsibility of pediatric audiologists. Reasons for failing to report vary.

DESIGN: The Early Hearing Detection and Intervention-Pediatric Audiology Links to Services (EHDI) facility survey was used to inform reporting compliance of audiology facilities throughout the United States. The survey was disseminated via articles, newsletters, and call-to-action notices to audiologists.

RESULTS: Among 1024 facilities surveyed, 88 (8.6%) reported that they did not report newborn's hearing findings to their state EHDI program. Not knowing how to report to the state EHDI program was the most frequently chosen reason (60%). However, among the 936 facilities that were compliant with the reporting requirements, 51 estimated that they reported less than two-third of all hearing evaluation results (5.4%). Some facilities did not report a normal-hearing result and some failed to report because they assumed another facility would report the hearing results.

CONCLUSIONS: Survey results indicated that audiologists were compliant reporting hearing results to the state EHDI programs. However, there is room for improvement. Regular provider outreach and training by the state EHDI program is necessary to ensure those who are not reporting will comply and to clarify reporting requirements for those who are already compliant.

Cicuto Ferreira Rocha NA, de Campos AC, Cicuto Ferreira Rocha F, Pereira Dos Santos Silva F.

Microcephaly and Zika virus: Neuroradiological aspects, clinical findings and a proposed framework for early evaluation of child development.

Infant Behav Dev. 2017 Jul 26;49:70-82. doi: 10.1016/j.infbeh.2017.07.002. [Epub ahead of print]

BACKGROUND AND AIMS: As the recent outbreak of microcephaly cases caused by Zika virus has been declared a global health emergency, providing assessment guidelines for multidisciplinary teams providing early developmental screening and stimulation to infants with microcephaly is much needed. Thus, the aim of this manuscript is to provide an overview on what is known about neuroradiological aspects and clinical findings in infants with microcephaly caused by Zika virus and to propose a framework for early evaluation of child development.

METHODS: The keywords "Zika virus" and "microcephaly" were searched in PubMed database for articles published from inception to May 2017. These texts were reviewed, and the ones addressing neuroradiological and clinical findings in infants were selected. Recommendations for early assessment were made based on the International Classification of Functionality Disability and Health (ICF) model.

OUTCOMES AND RESULTS: The database search yielded 599 publications and 36 were selected. The studies detected microcephaly with diffuse brain malformations and calcifications, ventriculomegaly, optic nerve hypoplasia, macular atrophy, cataracts, impaired visual and hearing function, arthrogryposis, spasticity, hyperreflexia, irritability, tremors, and seizures, but very little is known about early development. Early assessments were described based on the ICF domains (Body Function and Structures, Activities and Participation and Contextual factors).

CONCLUSION AND IMPLICATIONS: Studies published showed abnormal brain, optic, neurologic and orthopedic findings, but very little is known about other aspects of functioning in infants with microcephaly caused by Zika virus. The biopsychosocial model based on the ICF paradigm provides an adequate framework to describe the condition of the infant with microcephaly receiving rehabilitative efforts to minimize disability. Efforts towards early identification of developmental delays should be taken within the first six months of life.

Cunningham M, Thomson V, McKiever E, Dickinson LM, Furniss A, Allison MA.

Infant, maternal, and hospital factors' role in loss to follow-up after failed newborn hearing screening.

Acad Pediatr. 2017 May 23. pii: S1876-2859(17)30321-2. doi: 10.1016/j.acap.2017.05.005. [Epub ahead of print]

OBJECTIVE: Completion of newborn hearing screening (NBHS) is recommended by 1 month old. Delays and loss to follow-up and documentation (LTF/LTD) after failed NBHS are common. Committees of experts have established hospital guidelines to reduce LTF/LTD. We aimed to identify maternal and infant factors associated with LTF/LTD and determine if adherence to hospital guidelines is associated with timely completion of follow-up screening.

METHODS: We conducted a retrospective study of all infants born in Colorado hospitals who failed the newborn admission hearing screening from 2007 to 2012 and a cross-sectional survey of NBHS coordinators at Colorado birthing hospitals. Neonatal intensive care unit infants were excluded. Outcomes included documented completion of the follow-up NBHS and completion by 1 month. Data sources comprised the electronic birth record, infant hearing integrated data system, and NBHS coordinator survey. Data were analyzed by logistic regression.

RESULTS: A total of 13,904 newborns did not pass the newborn admission hearing screening from 2007 to 2012, and 11,422 (82%) had documentation of a completed follow-up screening. A total of 10,558 (76%) completed follow-up screening by 1 month. All 53 NBHS coordinators completed the survey. Maternal age, education, smoking, and birth country; and payer, race, birth order, and population density were associated with completion of follow-up hearing screening. Maternal education, payer, population density, birth weight, and cleft lip were associated with completion by 1 month of age. Only birth in a facility that charges a rescreening fee was associated with completion of follow-up screening.

CONCLUSIONS: Low-income, rural, and minority infants are at risk for LTF. Further studies are needed to determine if adherence to guidelines can overcome barriers to follow-up.

Dejaco D, Aregger FC, Hurth HV, Kegele J, Muigg V, Oberhammer L, Bunk S, Fischer N, Pinggera L, Riedl D, Otieno A, Agbenyega T, Adegnik AA, Riechelmann H, Lackner P, Zorowka P, Kreamsner P, Schmutzhard J.

Evaluation of transient-evoked otoacoustic emissions in a healthy 1 to 10 year pediatric

[cohort in Sub-Saharan Africa.](#)

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:65-69. doi: 10.1016/j.ijporl.2017.07.029. Epub 2017 Jul 24.

OBJECTIVE: Transient-evoked otoacoustic emissions (TEOAEs) monitor cochlear function. High pass rates have been reported for industrialized countries. Pass rates in low and middle income countries such as Sub-Saharan Africa are rare, essentially lower and available for children up to 4 years of age and frequently based on hospital recruitments. This study aims at providing additional TEOAE pass rates of a healthy Sub-Saharan cohort aged 1-10 years with data from Gabon, Ghana and Kenya. Potentially confounding factors (recruitment site, age) are taken into consideration.

METHODS: Healthy children were recruited in hospitals, schools and kindergartens. Inclusion criteria were age 1-10 years and normal otoscopic findings. Exclusion criteria were any sickness or physical ailment potentially impairing the hearing capacity. Five measurements per ear were performed with Capella Cochlear Emission Analyzer (MADSEN, Germany). An overall wave reproducibility of above 60% served as pass-criterion. Pass rates were compared between recruitment sites and age groups (1-5 and 6-10 years).

RESULTS: Overall pass rate was 87.5% (n = 264; 231 passes vs. 33 fails). Of these 84.0% of hospital recruited children passed (n = 156; 131 passes vs. 25 fails), compared to 92.6% of community recruitments (n = 108; 100 passes vs. 8 fails), which was significantly different p = 0.039). If analyzed by age groups, this difference was only observed in children younger than 6 years (p = 0.007).

CONCLUSION: Hospitals as recruitment sites for healthy controls seem to affect TEOAE pass rates. We advise for a cautious approach when recruiting healthy TEOAE control collectives under the age of 6 in a hospital setting. In children older than 6 years conventional pure-tone audiometry remains the standard method for hearing screening.

Dobbie AM.

[Evaluation and management of cytomegalovirus-associated congenital hearing loss.](#)

Curr Opin Otolaryngol Head Neck Surg. 2017 Oct;25(5):390-395. doi: 10.1097/MOO.0000000000000401.

PURPOSE OF REVIEW: The current article reviews the current literature related to congenital cytomegalovirus (CMV)-related hearing loss. The discussion will focus on the epidemiology, pathogenesis, and clinical presentation of human CMV infection as it pertains to hearing loss. Current methods of CMV diagnosis with a focus on the evolving trend toward broader neonatal screening protocols will also be explored. Discussion of medical, surgical, and audiologic management of the condition will also be addressed.

RECENT FINDINGS: Much of the current research on this topic is focused on improving detection of CMV through screening programs. Some advances in understanding cochlear pathogenesis have also been made.

SUMMARY: Congenital CMV infection remains an important cause of hearing loss in infants and children. Early detection of CMV infection can broaden treatment options and allow for improved hearing and language outcome for patients with CMV-associated sensorineural hearing loss.

Du Y, Huang L, Wang X, Cui Q, Cheng X, Zhao L, Ni T.

[Clinical data analysis of genotypes and phenotypes of deafness gene mutations in newborns: A retrospective study.](#)

Biosci Trends. 2017 Sep 12;11(4):460-468. doi: 10.5582/bst.2017.01070. Epub 2017 Jul 17.

We retrospectively analyzed newborns with deafness gene mutations and summarized the relationship between genotype and phenotype to provide a basis for genetic counseling. We studied 582 subjects positive for deafness gene mutations that were treated in the otology outpatient department of Beijing Tongren Hospital, Capital Medical University, between April 2012 and April 2016. The subjects were divided into 3 categories: a diagnosed group (group A), which was further subdivided into subgroups A1 (homozygous and compound heterozygous GJB2 mutations) and A2 (homozygous and compound heterozygous SLC26A4 mutations); a drug-induced deafness group (group B, mitochondrial (Mt) gene mutations); and a mutation carrier group (group C), which was further subdivided into the subgroups C1 (GJB2 heterozygous mutations), C2 (SLC26A4 heterozygous mutations), C3 (GJB3 heterozygous mutations), and C4 (double gene mutations). Partial sequences positive for GJB2 or SLC26A4 were sequenced and analyzed for mutations. Subjects underwent otoscopic examination and comprehensive audiological evaluation, and temporal bone computerized tomography and/or inner ear magnetic resonance imaging were performed. GJB2 235delC was the most common mutation locus. The highest proportion of deafness detected during universal newborn hearing screening was for drug-induced deafness, whereas the lowest was for the diagnosed group. GJB2 gene mutations mainly resulted in flat-type, profound-to-severe sensorineural hearing loss (SNHL). SLC26A4 gene mutation was mainly associated with high-frequency drop-type and profound-severe SNHL and was closely related to enlargement of the vestibular aqueduct.

Dudda R, Muniyappa HP, Puttaraju S, Lakshmi MS.

[A qualitative study on knowledge and attitude towards risk factors, early identification and intervention of infant hearing loss among puerperal mothers- a short survey.](#)

J Clin Diagn Res. 2017 Jul;11(7):MC01-MC05. doi: 10.7860/JCDR/2017/25837.10238. Epub 2017 Jul 1.

INTRODUCTION: Maternal active participation and their support are critical for the success of early hearing loss detection program. Erroneous maternal decisions may have large life long consequences on the infant's life. The mothers' knowledge and their attitudes towards infant hearing loss is the basis for their decisions.

AIM: The present study was done to determine the mothers' knowledge and their attitude towards risk factors of infant hearing loss, its early identification and intervention and also awareness of effect of consanguinity on hearing loss.

MATERIALS AND METHODS: In this cross-sectional questionnaire survey study, a total of 100 mothers were interviewed using the questionnaire which consisted of three sections namely risk factors, early identification and early intervention of hearing loss. Chi-square test was used to establish relationship between consanguineous and non-consanguineous mother's responses to its effect on hearing loss. A p-value < 0.05 was considered as significant.

RESULTS: Mothers' awareness was significantly high for visible causes (ear pain/discharge, head injury and slap to ear) of hearing loss. Positive attitude was seen for importance of screening programs and follow up testing. Moderate level of awareness was found on hazards of consanguinity and benefits of early identification. However, mothers were least aware of neonatal jaundice, NICU admission (>5 days), signs of late-onset and neural hearing loss, management of hearing loss, hearing aid fitting and therapy necessity, which might interfere in early detection and intervention of hearing loss.

CONCLUSION: It is crucial to educate mothers on few risk factors and management of hearing loss to reduce its consequences.

Fengler I, Nava E, Villwock AK, Büchner A, Lenarz T, Röder B.

[Multisensory emotion perception in congenitally, early, and late deaf CI users.](#)

PLoS One. 2017 Oct 12;12(10):e0185821. doi: 10.1371/journal.pone.0185821. eCollection 2017.

Emotions are commonly recognized by combining auditory and visual signals (i.e., vocal and facial expressions). Yet it is unknown whether the ability to link emotional signals across modalities depends on early experience with audio-visual stimuli. In the present study, we investigated the role of auditory experience at different stages of development for auditory, visual, and multisensory emotion recognition abilities in three groups of adolescent and adult cochlear implant (CI) users. CI users had a different deafness onset and were compared to three groups of age- and gender-matched hearing control participants. We hypothesized that congenitally deaf (CD) but not early deaf (ED) and late deaf (LD) CI users would show reduced multisensory interactions and a higher visual dominance in emotion perception than their hearing controls. The CD (n = 7), ED (deafness onset: <3 years of age; n = 7), and LD (deafness onset: >3 years; n = 13) CI users and the control participants performed an emotion recognition task with auditory, visual, and audio-visual emotionally congruent and incongruent nonsense speech stimuli. In different blocks, participants judged either the vocal (Voice task) or the facial expressions (Face task). In the Voice task, all three CI groups performed overall less efficiently than their respective controls and experienced higher interference from incongruent facial information. Furthermore, the ED CI users benefitted more than their controls from congruent faces and the CD CI users showed an analogous trend. In the Face task, recognition efficiency of the CI users and controls did not differ. Our results suggest that CI users acquire multisensory interactions to some degree, even after congenital deafness. When judging affective prosody they appear impaired and more strongly biased by concurrent facial information than typically hearing individuals. We speculate that limitations inherent to the CI contribute to these group differences.

Fitzpatrick EM, Dos Santos JC, Grandpierre V, Whittingham J.

Exploring reasons for late identification of children with early-onset hearing loss.

Int J Pediatr Otorhinolaryngol. 2017 Sep;100:160-167. doi:10.1016/j.ijporl.2017.06.039. Epub 2017 Jul 1.

INTRODUCTION: Several studies have shown that early identification of childhood hearing loss leads to better language outcomes. However, delays in the confirmation of hearing loss persist even in the presence of well-established universal newborn hearing screening programs (UNHS). The objective of this population-based study was to document the proportion of children who experienced delayed confirmation of congenital and early onset hearing loss in a UNHS program in one region of Canada. The study also sought to determine the reasons for delayed confirmation of hearing loss in children.

METHODS: Population level data related to age of first assessment, age of identification and clinical characteristics were collected prospectively for all children identified through the UNHS program. We documented the number of children who experienced delay (defined as more than 3 months) from initial audiologic assessment to confirmation of hearing loss. A detailed chart review was subsequently performed to examine the reasons for delay to confirmation.

RESULTS: Of 418 children identified from 2003 to 2013, 182 (43.5%) presented with congenital or early onset hearing loss, of whom 30 (16.5%) experienced more than 3 months delay from initial audiologic assessment to confirmation of their hearing disorder. The median age of first assessment and confirmation of hearing loss for these 30 children was 3.7 months (IQR: 2.0, 7.6) and 13.8 months (IQR: 9.7, 26.1) respectively. Close examination of the factors related to delay to confirmation revealed that for the overwhelming majority of children, a constellation of factors contributed to late diagnosis. Several children (n = 22; 73.3%) presented with developmental/medical issues, 15 of whom also had middle ear dysfunction at assessment, and 9 of whom had documented family follow-up concerns. For the remaining eight children, additional reasons included ongoing middle ear dysfunction for five children, complicated by family follow-up concerns (n = 3) and mild hearing loss (n = 1) and the remaining three children had isolated reasons related to family follow-up (n = 1) or mild hearing loss (n = 2).

CONCLUSION: Despite the progress made in the early detection of pediatric hearing loss since UNHS, a substantial number of children referred for early assessment can experience late confirmation and intervention. In particular, infants with developmental and/or medical issues including middle ear disorders are at particular risk for longer time to confirmation of hearing loss.

Fitzpatrick EM, Al-Essa RS, Whittingham J, Fitzpatrick J.

Characteristics of children with unilateral hearing loss.

Int J Audiol. 2017 Nov;56(11):819-828. doi: 10.1080/14992027.2017.1337938. Epub 2017 Jun 22.

OBJECTIVE: The purpose of this study was to describe the clinical characteristics of children with unilateral hearing loss (UHL), examine deterioration in hearing, and explore amplification decisions.

DESIGN: Population-based data were collected prospectively from time of diagnosis. Serial audiograms and amplification details were retrospectively extracted from clinical charts to document the trajectory and management of hearing loss.

SAMPLE: The study included all children identified with UHL in one region of Canada over a 13-year period (2003-2015) after implementation of universal newborn hearing screening.

RESULTS: Of 537 children with permanent hearing loss, 20.1% (108) presented with UHL at diagnosis. They were identified at a median age of 13.9 months (IQR: 2.8, 49.0). Children with congenital loss were identified at 2.8 months (IQR: 2.0, 3.6) and made up 47.2% (n=51), reflecting that a substantial portion had late-onset, acquired or late-identified loss. A total of 42.4% (n=39) showed deterioration in hearing, including 16 (17.4%) who developed bilateral loss. By study end, 73.1% (79/108) of children had received amplification recommendations.

CONCLUSIONS: Up to 20% of children with permanent HL are first diagnosed with UHL. About 40% are at risk for deterioration in hearing either in the impaired ear and/or in the normal hearing ear.

Fontenot TE, Giardina CK, Teagle HF, Park LR, Adunka OF, Buchman CA, Brown KD, Fitzpatrick DC.

Clinical role of electrocochleography in children with auditory neuropathy spectrum disorder.

Int J Pediatr Otorhinolaryngol. 2017 Aug;99:120-127. doi:10.1016/j.ijporl.2017.05.026. Epub 2017 Jun 5.

OBJECTIVES: To assess electrocochleography (ECoChG) to tones as an instrument to account for CI speech perception outcomes in children with auditory neuropathy spectrum disorder (ANSD).

MATERIALS & METHODS: Children (<18 years) receiving CIs for ANSD (n = 30) and non-ANSD (n = 74) etiologies of hearing loss were evaluated with ECoChG using tone bursts (0.25-4 kHz). The total response (TR) is the sum of spectral peaks of responses across frequencies. The compound action potential (CAP) and the auditory nerve neurophonic (ANN) in ECoChG waveforms were used to estimate nerve activity and calculate nerve score. Performance on open-set monosyllabic word tests was the outcome measure. Standard statistical methods were applied.

RESULTS: On average, TR was larger in ANSD than in non-ANSD subjects. Most ANSD (73.3%) and non-ANSD (87.8%) subjects achieved open-set speech perception; TR accounted for 33% and 20% of variability in the outcomes, respectively. In the ANSD group, the PTA accounted for 69.3% of the variability, but there was no relationship with outcomes in the non-ANSD group. In both populations, nerve score was sensitive in identifying subjects at risk for not acquiring open-set speech perception, while the CAP and the ANN were more specific.

CONCLUSION: In both subject groups, the TRs correlated with outcomes but these measures were notably larger in the ANSD group. There was also strong correlation between PTA and speech perception outcome in ANSD group. In both subject populations, weaker evidence of neural activity was related to failure to achieve open-set speech perception.

Franck C, Vorwerk W, Köhn A, Rißmann A, Vorwerk U.

Prevalence, risk factors and diagnostics of hearing impairment in preterm infants.

[Article in German; Abstract available in German from the publisher]

Laryngorhinootologie. 2017 Jun;96(6):354-360. doi: 10.1055/s-0043-109512. Epub 2017 Jul 11.

INTRODUCTION: The preterm birth is clearly associated with increased risk of developing congenital hearing impairment. Therefore, special attention must be paid to the postnatal control of auditory function in all preterm infants. The present work investigates if the latest scientific findings regarding prevalence, clinical diagnostics, therapy and risk factors of hearing impairment in premature infants are regularly implemented in daily practice.

METHODS: At the department of phoniatrics and pediatric audiology of the University of Magdeburg, the treatment data of 126 preterm children born between 2006 and 2011 were evaluated retrospectively. The additional analysis of all records available at the screening center (n=67 640) covering this period enables drawing conclusions on the total number and prevalence of hearing impairment in preterm infants in Saxony-Anhalt.

RESULTS: Almost all premature babies, like mature newborns, underwent postnatal hearing screening of both ears. The data analysis shows that the practical implementation often does not comply with the guideline of the G-BA (Gemeinsamer Bundesausschuss) in all details. For example, the recommended screening method for preterm infants (AABR) or the screening and treatment timing are not always applied in accordance with the guidelines of the G-BA.

DISCUSSION: Assessment of the practical implementation of universal newborn hearing screening was planned at the time of the introduction of the hearing screening program by the G-BA. As a part of this investigation, the practical care of vulnerable groups such as preterm infants must be given special attention. Based on the collected data, the diagnostics and therapy should be unified. Regardless of the maternity clinic where the infants were born, there should be the same opportunity for early diagnosis and thus for prognostically better treatment of congenital hearing impairment. Rapid postnatal fitting with hearing aid can stimulate the maturation of the central auditory system and potentially help to avoid problems of hearing and speech development.

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

Monitoring neonates for ototoxicity.

Int J Audiol. 2017 Jun 22:1-8. doi: 10.1080/14992027.2017.1339130. [Epub ahead of print]

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.

Garinis AC, Liao S, Cross CP, Galati J, Middaugh JL, Mace JC, Wood AM, McEvoy L, Moneta L, Lubianski T, Coopersmith N, Vigo N, Hart C, Riddle A, Ettinger O, Nold C, Durham H, MacArthur C, McEvoy C, Steyger PS.

Effect of gentamicin and levels of ambient sound on hearing screening outcomes in the neonatal intensive care unit: A pilot study.

Int J Pediatr Otorhinolaryngol. 2017 Jun;97:42-50. doi: 10.1016/j.ijporl.2017.03.025. Epub

OBJECTIVE: Hearing loss rates in infants admitted to neonatal intensive care units (NICU) run at 2-15%, compared to 0.3% in full-term births.

The etiology of this difference remains poorly understood. We examined whether the level of ambient sound and/or cumulative gentamicin (an aminoglycoside) exposure affect NICU hearing screening results, as either exposure can cause acquired, permanent hearing loss. We hypothesized that higher levels of ambient sound in the NICU, and/or gentamicin dosing, increase the risk of referral on the distortion product otoacoustic emission (DPOAE) assessments and/or automated auditory brainstem response (AABR) screens.

METHODS: This was a prospective pilot outcomes study of 82 infants (<37 weeks gestational age) admitted to the NICU at Oregon Health & Science University. An ER-200D sound pressure level dosimeter was used to collect daily sound exposure in the NICU for each neonate. Gentamicin dosing was also calculated for each infant, including the total daily dose based on body mass (mg/kg/day), as well as the total number of treatment days.

DPOAE and AABR assessments were conducted prior to discharge to evaluate hearing status. Exclusion criteria included congenital infections associated with hearing loss, and congenital craniofacial or otologic abnormalities.

RESULTS: The mean level of ambient sound was 62.9 dBA (range 51.8-70.6 dBA), greatly exceeding American Academy of Pediatrics (AAP) recommendation of <45.0 dBA. More than 80% of subjects received gentamicin treatment. The referral rate for (i) AABRs, (frequency range: 1000-4000 Hz), was 5%; (ii) DPOAEs with a broad F2 frequency range (2063-10031 Hz) was 39%; (iii) DPOAEs with a low-frequency F2 range (<4172 Hz) was 29%, and (iv) DPOAEs with a high-frequency F2 range (>4172 Hz) was 44%. DPOAE referrals were significantly greater for infants receiving >2 days of gentamicin dosing compared to fewer doses (p = 0.004). The effect of sound exposure and gentamicin treatment on hearing could not be determined due to the low number of NICU infants without gentamicin exposure (for control comparisons).

CONCLUSION: All infants were exposed to higher levels of ambient sound that substantially exceed AAP guidelines. More referrals were generated by DPOAE assessments than with AABR screens, with significantly more DPOAE referrals with a high-frequency F2 range, consistent with sound- and/or gentamicin-induced cochlear dysfunction. Adding higher frequency DPOAE assessments to existing NICU hearing screening protocols could better identify infants at-risk for ototoxicity.

Geers AE, Mitchell CM, Warner-Czyz A, Wang NY, Eisenberg LS; CDaCI Investigative Team.

Early sign language exposure and cochlear implantation benefits.

Pediatrics. 2017 Jul;140(1). pii: e20163489. doi: 10.1542/peds.2016-3489. Epub 2017 Jun 12.

BACKGROUND: Most children with hearing loss who receive cochlear implants (CI) learn spoken language, and parents must choose early on whether to use sign language to accompany speech at home. We address whether parents' use of sign language before and after CI positively influences auditory-only speech recognition, speech intelligibility, spoken language, and reading outcomes.

METHODS: Three groups of children with CIs from a nationwide database who differed in the duration of early sign language exposure provided in their homes were compared in their progress through elementary grades. The groups did not differ in demographic, auditory, or linguistic

characteristics before implantation.

RESULTS: Children without early sign language exposure achieved better speech recognition skills over the first 3 years postimplant and exhibited a statistically significant advantage in spoken language and reading near the end of elementary grades over children exposed to sign language. Over 70% of children without sign language exposure achieved age-appropriate spoken language compared with only 39% of those exposed for 3 or more years. Early speech perception predicted speech intelligibility in middle elementary grades. Children without sign language exposure produced speech that was more intelligible (mean = 70%) than those exposed to sign language (mean = 51%).

CONCLUSIONS: This study provides the most compelling support yet available in CI literature for the benefits of spoken language input for promoting verbal development in children implanted by 3 years of age. Contrary to earlier published assertions, there was no advantage to parents' use of sign language either before or after CI.

Gouws N, Swanepoel W, De Jager LB.

Wideband acoustic immittance for assessing middle ear functioning for preterm neonates in the neonatal intensive care unit.

S Afr J Commun Disord. 2017 Jun 28;64(1):e1-e11. doi: 10.4102/sajcd.v64i1.182.

BACKGROUND: The primary aim of newborn hearing screening is to detect permanent hearing loss. Because otoacoustic emissions (OAEs) and automated auditory brainstem response (AABR) are sensitive to hearing loss, they are often used as screening tools. On the other hand, false-positive results are most often because of transient outer- and middle ear conditions. Wideband acoustic immittance (WAI), which includes physical measures known as reflectance and absorbance, has shown potential for accurate assessment of middle ear function in young infants.

OBJECTIVE: The main objective of this study was to determine the feasibility of WAI as a diagnostic tool for assessing middle ear functioning in preterm neonates in the neonatal intensive care unit (NICU) designed for premature and ill neonates. A further objective was to indicate the difference between the reflectance values of tones and click stimuli.

METHOD: Fifty-six at-risk neonates (30 male and 26 female), with a mean age at testing of 35.6 weeks (range: 32-37 weeks) and a standard deviation of 1.6 from three private hospitals, who passed both the distortion product otoacoustic emission (DPOAE) and AABR tests, were evaluated prior to discharge from the NICU. Neonates who presented with abnormal DPOAE and AABR results were excluded from the study. WAI was measured by using chirp and tone stimuli. In addition to reflectance, the reflectance area index (RAI) values were calculated.

RESULTS: Both tone and chirp stimuli indicated high-power reflectance values below a frequency of 1.5 kHz. Median reflectance reached a minimum of 0.67 at 1 kHz - 2 kHz but increased to 0.7 below 1 kHz and 0.72 above 2 kHz for the tone stimuli. For chirp stimuli, the median reflectance reached a minimum of 0.51 at 1 kHz - 2 kHz but increased to 0.68 below 1 kHz and decreased to 0.5 above 2 kHz. A comparison between the present study and previous studies on WAI indicated a substantial variability across all frequency ranges.

CONCLUSION: These WAI measurements conducted on at-risk preterm NICU neonates (mean age at testing: 35.6 weeks, range: 32-37 weeks) identified WAI patterns not previously reported in the literature. High reflective values were obtained across all frequency ranges. The age of the neonates when tested might have influenced the results. The neonates included in the present study were very young preterm neonates compared to the ages of neonates in previous studies. WAI measured in at-risk preterm neonates in the NICU was variable with environmental and internal noise influences. Transient conditions affecting the sound-conduction pathway might have influenced the results. Additional research is required to investigate WAI testing in ears with and without middle ear dysfunction. The findings of the current study imply that in preterm neonates it was not possible to determine the feasibility of WAI as a diagnostic tool to differentiate between ears with and without middle ear pathology.

Govender SM, Khan NB.

Knowledge and cultural beliefs of mothers regarding the risk factors of infant hearing loss and awareness of audiology services.

J Public Health Afr. 2017 Sep 4;8(1):557. doi: 10.4081/jphia.2017.557.eCollection 2017 Jun 23.

The aim of the paper is to describe the knowledge of mothers in Durban, South Africa, regarding risk factors of hearing loss in infants and their awareness of audiology services, and to describe their cultural beliefs about the risk factors for hearing loss in infants. A descriptive survey design with quantitative **METHODS** of analysis were used. Conveniently sampled mothers (n=102) receiving postnatal care for their infants from eight provincial clinics within Durban consented to participate, yielding a response rate of 48%. A questionnaire was used to collect the data and the Cronbach α was calculated yielding a score of 0.835, indicating good internal consistency and reliability of the questionnaire. Sixty percent of the mothers were aware of risk factors, such as middle ear infections, ototoxic medication and consumption of alcohol during pregnancy. Seventy percent were unaware that NICU/mechanical ventilation for more than 5 days, prematurity, rubella and jaundice are considered risk factors for hearing loss, implying a need to create awareness amongst mothers regarding such risk factors. Sixty percent (n=62) believed that bewitchment and ancestral curses can cause hearing loss. Cultural beliefs were associated with hearing loss, therefore, health professionals need to demonstrate cultural competence when providing audiology services, especially in a culturally and linguistically diverse countries such as South Africa. Although the mothers had an average knowledge about risk factors, two thirds did not know which professional to seek help from. There is a need to create awareness amongst mothers regarding the risk factors of infant hearing loss as well as audiology services in order to facilitate early detection and intervention. There is a need for health professionals to demonstrate cultural competence when working with their patients.

Grandori F, Hayes D.

Reflections on Lake Como Conferences (2000-2016).

Am J Audiol. 2017 Oct 12;26(3S):467-468. doi:10.1044/2017_AJA-17-0062.

PURPOSE: We present an overview of the conceptualization and development of the Newborn Hearing Screening and Hearing Across the Lifespan (Lake Como) conferences from 2000 to 2016.

Grosse SD, Riehle-Colarusso T, Gaffney M, Mason CA, Shapira SK, Sontag MK, Braun KVN, Iskander J.

CDC grand rounds: Newborn screening for hearing loss and critical congenital heart disease.

MMWR Morb Mortal Wkly Rep. 2017 Aug 25;66(33):888-890. doi:10.15585/mmwr.mm6633a4.

Newborn screening is a public health program that benefits 4 million U.S. infants every year by enabling early detection of serious conditions, thus affording the opportunity for timely intervention to optimize outcomes (1). States and other U.S. jurisdictions decide whether and how to regulate newborn screening practices. Most newborn screening is done through laboratory analyses of dried bloodspot specimens collected from newborns. Point-of-care newborn screening is typically performed before discharge from the birthing facility. The Recommended Uniform Screening Panel includes two point-of-care conditions for newborn screening: hearing loss and critical congenital heart disease (CCHD). The objectives of point-of-care screening for these two conditions are early identification and intervention to improve neurodevelopment, most notably language and related skills among infants with permanent hearing loss, and to prevent death or severe disability resulting from delayed diagnosis of CCHD. Universal screening for hearing loss using otoacoustic emissions or automated auditory brainstem response was endorsed by the Joint Committee on Infant Hearing in 2000 and 2007* and was incorporated in the first Recommended Uniform Screening Panel in 2005. Screening for CCHD using pulse oximetry was recommended by the Advisory Committee on Heritable Disorders in Newborns and Children in 2010 based on an evidence review(†) and was added to the Recommended Uniform Screening Panel in 2011.(§).

Guerzoni L, Cuda D.

[Speech processor data logging helps in predicting early linguistic outcomes in implanted children.](#)

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:81-86. doi: 10.1016/j.ijporl.2017.07.026. Epub 2017 Jul 25.

OBJECTIVE: To analyse the value of listening-data logged in the speech processor on the prediction of the early auditory and linguistic skills in children who received a cochlear implant in their first 2 years of life.

STUDY DESIGN: Prospective observational non-randomized study.

METHODS: Ten children with profound congenital sensorineural hearing loss were included in the study. The mean age at CI activation was 16.9 months (SD ± 7.2; range 10-24). The auditory skills were evaluated with the Infant Toddler Meaningful Inventory Scale and the Category of Auditory Performance. Lexical level was assessed with the MacArthur-Bates Communicative Development Inventory. The overall data of average daily use and acoustic scene-analyses were extracted from Data Logging system. The effect of the one-year cumulative listening time to speech (in quiet) and speech-in-noise on the auditory and lexical scores was analysed.

RESULTS: A significant positive correlation was found between speech in quiet exposure time at low loudness level (<70 dB) and lexical quotient after one year of CI use. Infant Toddler Meaningful Inventory Scale was negatively correlated with the highest speech-in-noise loudness levels (>80 dB). The Category of Auditory Performance was not related to the logged data.

CONCLUSION: The listening environment can influence the early functional outcomes in younger implanted children. In this perspective, the data logging system is a promising tool in predicting early linguistic and auditory outcomes.

Hall WC.

[What you don't know can hurt you: The risk of language deprivation by impairing sign language development in deaf children.](#)

Matern Child Health J. 2017 May;21(5):961-965. doi: 10.1007/s10995-017-2287-y.

A long-standing belief is that sign language interferes with spoken language development in deaf children, despite a chronic lack of evidence supporting this belief. This deserves discussion as poor life outcomes continue to be seen in the deaf population. This commentary synthesizes research outcomes with signing and non-signing children and highlights fully accessible language as a protective factor for healthy development. Brain changes associated with language deprivation may be misrepresented as sign language interfering with spoken language outcomes of cochlear implants. This may lead to professionals and organizations advocating for preventing sign language exposure before implantation and spreading misinformation. The existence of one-time-sensitive-language acquisition window means a strong possibility of permanent brain changes when spoken language is not fully accessible to the deaf child and sign language exposure is delayed, as is often standard practice. There is no empirical evidence for the harm of sign language exposure but there is some evidence for its benefits, and there is growing evidence that lack of language access has negative implications. This includes cognitive delays, mental health difficulties, lower quality of life, higher trauma, and limited health literacy. Claims of cochlear implant- and spoken language-only approaches being more effective than sign language-inclusive approaches are not empirically supported. Cochlear implants are an unreliable standalone first-language intervention for deaf children. Priorities of deaf child development should focus on healthy growth of all developmental domains through a fully-accessible first language foundation such as sign language, rather than auditory deprivation and speech skills.

Han R, Li L, Duan L, Xia Y, Kuyaxi P, Zhao J, Zhao Q, Zhang H, Chen Y.

[Efficiency of microarray and SNPscan for the detection of hearing loss gene in 71 cases with nonsyndromic hearing loss.](#)

Medicine (Baltimore). 2017 Jun;96(25):e7149. doi: 10.1097/MD.0000000000007149.

We aim to screen the mutations of 3 hearing loss (HL) genes (GJB2, SLC26A4, and 12S rRNA) in 71 cases with nonsyndromic hearing loss (NSHL) using microarray and SNPscan, and identify the roles of nonhotspot mutation of these genes in the screening of NSHL. Seventy-one cases with moderate or severe neurosensory deafness confirmed in our department from July 2014 to December 2015 including 25 Uyghur minorities and 46 Han Chinese were included in this study. The type of mutations in GJB2, SLC26A4, and 12S rRNA genes were detected using microarray and SNPscan, respectively. Statistical difference was noticed in the detection rate of the HL genes in 71 cases. Using microarray, deafness genes were identified in 10 subjects (14.08%), while 22 cases (30.98%) were confirmed with the presence of deafness genes using the SNPscan. Compared with the microarray, remarkable difference was noticed in the detection rate of SNPscan (P<.05). Nonhotspot mutation in GJB2, SLC26A4, and 12S rRNA genes played a crucial role in the pathogenesis of NSHL. SNPscan contributed to elevation of detection rate of NSHL in clinical practice.

Harris MS, Dodson EE.

[Hearing health access in developing countries.](#)

Curr Opin Otolaryngol Head Neck Surg. 2017 Oct;25(5):353-358. doi:10.1097/MOO.0000000000000392.

PURPOSE OF REVIEW: The developing world carries a disproportionate burden of hearing loss. Individuals with hearing loss in austere settings worldwide are also potentially impacted by their impairment to a greater extent owing to underdeveloped or nonexistent hearing health infrastructure. The purpose of this review is to examine the state of the literature on hearing health access in developing countries and identify areas for improvement.

RECENT FINDINGS: Over the last 10 years progress has been made in some areas, whereas other aspects of hearing health in developing countries have changed very little. There has been expansion of efforts to train primary care and local hearing healthcare providers to recognize and appropriately treat preventable causes of hearing loss in the developing world. Applications of telehealth to connect providers and patients in rural

locales have grown. Adaptions of newborn hearing screening programs that better fit local resources and customs have been reported. There has been a considerable increase in interest, including cost-benefit analyses, with regard to the use of cochlear implants in the developing world. **SUMMARY:** In spite of progress, the developing world still shoulders a disproportionate amount of the world's hearing loss, in particular chronic ear disease, and there is a paucity of well trained local hearing healthcare professionals to deal with the challenge. The role of international humanitarian efforts, telemedicine, and education is highlighted.

Hoey AW, Pai I, Driver S, Connor S, Wraige E, Jiang D.

Management and outcomes of cochlear implantation in patients with congenital cytomegalovirus (cCMV)-related deafness.

Cochlear Implants Int. 2017 Jul;18(4):216-225. doi:10.1080/14670100.2017.1315510. Epub 2017 May 9.

OBJECTIVE: Congenital Cytomegalovirus (cCMV) is a well-defined cause for neonatal mortality and morbidity, particularly sensorineural hearing loss and other neurodevelopmental disruption. We present a retrospective study which provides an overview of the assessment and preoperative work-up for patients diagnosed with cCMV and their cochlear implant (CI) outcomes.

METHOD: This was a retrospective case series study of all children with a confirmed diagnosis of cCMV who underwent cochlear implantation at St Thomas' Hospital from 2003 to 2015. Data were collected on the preoperative audiology, imaging findings, and neurological assessment. CI outcomes were measured using the Speech Intelligibility Rating (SIR), Category of Auditory Performance (CAP), and Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS).

RESULTS: Eleven patients underwent cochlear implantation, 45% had severe-to-profound hearing loss, and 55% had bilateral profound hearing loss. The mean age at initial assessment was 2.1 years (median 1.7, range 0.6-7.5) and the mean age of implantation was 4.0 years (median 2.5, range 0.9-11.8). The mean length of follow-up was 4.8 years (median 2.3, range 1.5-14). Six patients had bilateral simultaneous implantation (55%), four bilateral sequential (36%), and one unilateral (9%). Nine patients had white matter changes on magnetic resonance imaging, largely in the periventricular and cortical regions. Of the 11 patients, 4 (36%) had associated neurological comorbidities and 3 (27%) had additional neurocognitive developmental delay of varying severity. The majority of patients showed improvement in auditory outcomes. No statistically significant correlation was found between age of implantation, neurocognitive, and neurological comorbidities or length of follow-up and hearing outcomes.

CONCLUSION: While the overall outcomes were mixed, most children in our cohort were found to benefit from cochlear implantation. Our data also highlight the significant neurodevelopmental comorbidities associated with cCMV and their negative impact on CI outcomes. With the recent advances in medical treatment, this underlines the importance of multidisciplinary management of these patients.

Kasakura-Kimura N, Masuda M, Mutai H, Masuda S, Morimoto N, Ogahara N, Misawa H, Sakamoto H, Saito K, Matsunaga T.

WFS1 and GJB2 mutations in patients with bilateral low-frequency sensorineural hearing loss.

Laryngoscope. 2017 Sep;127(9):E324-E329. doi: 10.1002/lary.26528. Epub 2017 Mar8.

OBJECTIVE: Evaluating the prevalence of specific gene mutations associated with a certain audiometric configuration facilitates clinical assessment of patients with sensorineural hearing loss (SNHL). WFS1 is responsible for autosomal dominant nonsyndromic deafness 6/14/38 and is the most frequent genetic cause of low-frequency SNHL (LFSNHL); however, the exact prevalence of WFS1 mutations in LFSNHL is unknown. Therefore, we evaluated genetic mutations and clinical features in patients with nonsyndromic bilateral LFSNHL, focusing on the WFS1.

STUDY DESIGN: Retrospective case series from 2002 to 2013 at the National Hospital Organization Tokyo Medical Center and collaborating hospitals.

METHODS: WFS1, GJB2, and mitochondrial DNA mutation screening was carried out for 74 of 1,007 Japanese probands with bilateral LFSNHL.

RESULTS: WFS1 and GJB2 mutations were identified in eight of 74 cases (10.8%). Four cases had heterozygous WFS1 mutations; one case had a heterozygous WFS1 mutation and a heterozygous GJB2 mutation; and three cases had biallelic GJB2 mutations. Three cases with WFS1 mutations were sporadic; two of them were confirmed to be caused by a de novo mutation based on the genetic analysis of their parents. In the case with mutations in both WFS1 and GJB2, a de novo mutation of WFS1 was confirmed in the proband's mother by genetic screening of the mother's parents.

CONCLUSION: Genetic screening focusing on LFSNHL has not been conducted. The present study first revealed the prevalence of specific gene mutations. WFS1 autosomal dominant mutations were identified even in sporadic cases. Our results also suggested a mutational hotspot in WFS1.

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. 2017 May 22:1-6. doi: 10.1080/14767058.2017.1326902. [Epub ahead of print]

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.

Kim SH, Choi BY, Park J, Jung EY, Cho SH, Park KH.

Maternal and placental factors associated with congenital hearing loss in very preterm neonates.

Pediatr Neonato. 2017 Jun;58(3):236-244. doi: 10.1016/j.pedneo.2016.05.003. Epub 2016 Aug 9.

BACKGROUND: Sensorineural hearing loss (SNHL) is a multifactorial disease that more frequently affects preterm newborns. Although a number

of maternal conditions have been reported to be associated with preterm birth, little information is available concerning maternal risk factors for the development of SNHL. We aimed to identify maternal and placental risk factors associated with a "refer" result on the newborn hearing screening (NHS) test and subsequently confirmed SNHL in very preterm neonates.

METHODS: This retrospective cohort study included 267 singleton neonates who were born alive after ≤ 32 weeks. Histopathologic examination of the placenta was performed, and clinical data were retrieved from a computerized perinatal database. Cases with two abnormal findings, "refer" on the NHS test, and presence of SNHL on the confirmation test were retrospectively reviewed based on electronic medical records.

RESULTS: Forty-two neonates (15.7%) showed a "refer" result, and, on the confirmation test, permanent SNHL was identified in 1.87% (5/267) of all neonates. Multivariate regression analysis revealed that the presence of funisitis was independently associated with a "refer" on the NHS test, whereas use of antenatal corticosteroids was statistically significantly associated with a reduced incidence of "refer" on the screening test. Neither histologic chorioamnionitis nor prematurity (as defined by low gestational age and birth weight) was associated with a "refer" on the NHS test. By contrast, multivariate analysis with occurrence of SNHL as a dependent variable identified no significant associations with the parameters studied, probably owing to the small total number of neonates with permanent SNHL.

CONCLUSION: Presence of funisitis was significantly and independently associated with increased risk of abnormal NHS results, while administration of antenatal corticosteroids was related to a normal NHS result. These findings support the hypothesis that a systemic fetal inflammatory response, manifested as funisitis, might play a role in the pathogenesis of SNHL in preterm neonates.

Kim SH, Lim JH, Han JJ, Jin YJ, Kim SK, Kim JY, Song JJ, Choi BY, Koo JW.

[Outcomes and limitations of hospital-based newborn hearing screening.](#)

Int J Pediatr Otorhinolaryngol. 2017 Jul;98:53-58. doi:10.1016/j.ijporl.2017.04.030. Epub 2017 Apr 20.

OBJECTIVES: Globally, newborn hearing screening (NHS) is variably incorporated into national healthcare systems. The authors reviewed the set-up and evolution process of a hospital-based NHS program in South Korea, where screening costs for low-income families are paid by the National Health Authority.

METHODS: The NHS process for 13805 newborns delivered in a tertiary referral center of South Korea from 2005 through 2014 was reviewed. Hearing screening was conducted using automated auditory brainstem response (AABR); hearing loss was confirmed by auditory brainstem response for newborns who did not pass the screening test.

RESULTS: The mean screening rate for hearing loss was 53.6% (7403 of 13805 newborns), which plateaued at 79.6% over time. Of the 14806 ears (7403 newborns), 1030 (7.0%) were assessed as "refer" on the first AABR, with 204 (1.4%) being assessed as "refer" on the second AABR. In hearing confirmation tests, 74 infants (1.0% of 7403 newborns) were diagnosed with hearing loss, including 13 infants (0.2%) with bilateral moderate to profound sensorineural hearing loss (SNHL). Hearing rehabilitation with long-term follow-up was confirmed in 11 infants.

CONCLUSIONS: In this hospital-based NHS program, the screening rate plateaued at $\square 50\%$ when the National Health Authority was not involved, but increased to $\square 70\%$ when the cost for low-income families was covered by the government. Among infants needing active hearing rehabilitation due to bilateral moderate to profound SNHL, 15% were lost to follow-up. These results demonstrate the need for a universal, mandatory NHS program to systematically register hearing-impaired infants within the government-sponsored public healthcare system.

Komínek P, Chrobok V, Zeleník K, Dršata J.

[\[Newborn hearing screening - importance, current state in the Czech Republic\]. \[Article in Czech\]](#)

Cas Lek Cesk. 2017 Summer;156(4):173-177.

The importance of early detection of hearing impairment in newborns and children and the early rehabilitation of hearing disorder with hearing aid or cochlear implant was demonstrated in a number of papers. As a result, newborn hearing screening was introduced in many countries around the world. The incidence of congenital hearing impairment has been underestimated for a long time, empirically determined incidence was 1:1000 neonates. Thanks to newborn hearing screening was revealed that incidence of congenital hearing impairment is 3 times higher. One out of 300 newborns has profound hearing loss (deafness) and 1 in 300 newborns has mild to moderate hearing loss. Moreover, in 1 out of 300 children hearing impairment would develop before the age of 18. In the Czech Republic, the screening of all newborns is still systematic and multilevel conducted in only three regions (Moravskoslezský, Královéhradecký, Pardubický). In these regions, statistics as well as assessments of individual stages of screening are carried out. In other regions, records of the number of screened children, number of rescreened children and detailed statistics of hearing impairment is missing. The authors summarize the basic information about the importance of screening, the history of screening and its organization. Provided information is based on experience with the gradual introduction of screening over recent years in the regions in which they work. Newborn hearing screening should be organized in several stages (1 - screening at maternity hospitals, 2 - rescreening on collaborating otolaryngology/phoniatric workplaces and 3 - detailed hearing examination in centers in children whose screening was negative). The authors focus on problems related to the organization of screening and offer practical advice (e.g. implementation of screening coordinators). They consider it is essential to have statistical evidence of examination at all levels. Introducing of newborn hearing screening is not a short-term task but a long-term (many years) challenge.

Koyano S, Morioka I, Oka A, Moriuchi H, Asano K, Ito Y, Yoshikawa T, Yamada H, Suzutani T, Inoue N; Japanese Congenital Cytomegalovirus Study Group.

[More than two years follow-up of infants with congenital cytomegalovirus infection in Japan.](#)

Pediatr Int. 2017 Oct 15. doi:10.1111/ped.13433. [Epub ahead of print]

BACKGROUND: The aim of the study was to evaluate outcomes of congenital cytomegalovirus (CMV) cases identified by our urine-filter screening assay after follow-up for more than 2 years, and to observe the clinical outcomes after anti-CMV therapies.

METHODS: Sixty of the 72 congenital CMV cases were enrolled. Forty-three cases asymptomatic at birth, 7 cases symptomatic at birth but untreated with anti-CMV drugs, and 10 symptomatic cases treated with anti-CMV drugs were clinically observed for more than 2 years.

RESULTS: Among the 43 asymptomatic cases, 3 cases developed hearing loss or language disabilities for which association with congenital CMV has been repeatedly reported and 2 cases demonstrated neurological sequelae of which etiology was unclear, indicating that the rate of CMV-associated late-onset sequelae was 7~12%. All 7 symptomatic infants without treatment developed sequelae, while 3 of the 10 treated cases were free from any sequelae.

CONCLUSIONS: The rate of late-onset sequelae observed in Japan is similar to that reported for cases in the US and European countries. The treatment of symptomatic cases with antiviral agents results in favorable clinical outcomes. Thus, newborn screening of congenital CMV infection by the collection of urine on filter paper is warranted.

Kubba H, Smyth A, Wong SC, Mason A.

[Ear health and hearing surveillance in girls and women with Turner's syndrome: recommendations from the Turner's Syndrome Support Society.](#)

Clin Otolaryngol. 2017 Jun;42(3):503-507. doi: 10.1111/coa.12750. Epub 2016 Sep28.

BACKGROUND: Turner's syndrome (TS) is a common chromosomal disorder, affecting one in 2000 newborn girls, in which part or all of one X chromosome is missing. Ear and hearing problems are very common in girls and women with TS. The aim of this review was to review the published literature to suggest recommendations for otological health surveillance.

METHOD: A keyword search of Ovid Medline was performed for published literature on the subject and evidence rated according to the GRADE criteria.

RESULTS: Middle ear disorders are very common and persistent in girls and women with TS as are progressive sensorineural hearing loss and balance disorders.

CONCLUSIONS: Otolaryngologists should be aware of the high prevalence and challenging nature of all forms of ear disease in individuals with TS. Early intervention may offer benefits to health and education, and we advocate routine lifelong annual hearing screening in this group.

Kumar A, Gupta SC, Sinha VR.

Universal hearing screening in newborns using otoacoustic emissions and brainstem evoked response in Eastern Uttar Pradesh.

Indian J Otolaryngol Head Neck Surg. 2017 Sep;69(3):296-299. doi: 10.1007/s12070-017-1081-x. Epub 2017 Feb 3.

The objectives were to determine the incidence of hearing impairment in a standardized population of neonates and to determine the significance of association of epidemiological and risk factors with neonatal hearing loss. A cohort of 600 newborns was selected for study and divided into two groups-525 in 'No Risk' group and remaining 75 in 'At Risk' group. The study protocol was carried out in three steps: (a) Screening of Hearing Loss with TOAE, done from 36 h after birth to 28 days of life, (b) Re-screening of hearing loss in newborns (of 4-12 weeks of age), who were tested positive for hearing loss in the first screening, done with DPOAE, (c) Confirmation of hearing loss with BERA, in those who were tested positive in both the first and second screening. In the study the incidence of hearing impairment in 600 infants screened was 6.67 per 1000 screened; 3.81 per 1000 screened in the Not at Risk group and 26.67 per 1000 screened in At Risk group. In At Risk group, admitted to the NICU, severe birth asphyxia and hyperbilirubinemia were found to be major risk factors. Loss to follow up was more in Not at Risk group and False Positive cases with TEOAE were more than DPOAE. BERA was found to be must for confirmation of hearing loss. Neonatal Hearing Screening of only At Risk population is likely to miss some hearing loss. Universal Hearing Screening should be the preferred strategy. Good follow up in the 'At Risk' group suggests that initial interventions in NICU had sensitized the parents for the possibility of hearing loss. This study recommends the introduction of two stage neonatal screening-rescreening protocol, using OAE and BERA, in the country in phased manner.

Li JN, Chen S, Zhai L, Han DY, Eshraghi AA, Feng Y, Yang SM, Liu XZ.

The advances in hearing rehabilitation and cochlear implants in China.

Ear Hear. 2017 May 3. doi: 10.1097/AUD.0000000000000441. [Epub ahead of print]

Hearing loss (HL) is a common sensory impairment in humans, with significant economic and social impacts. With nearly 20% of the world's population, China has focused on economic development and health awareness to improve the care for its hearing-impaired population. Recently, the Chinese government has initiated national programs such as the China Disabled Persons Federation to fund prevention, treatment, and rehabilitation of hearing impairment. Newborn hearing screening and auditory rehabilitation programs in China have expanded exponentially with government support. While facing many challenges and overcoming obstacles, cochlear implantation (CI) programs in China have also experienced considerable growth. This review discusses the implementation of CI programs for HL in China and presents current HL data including epidemiology, newborn hearing screening, and determination of genetic etiologies. Sharing the experience in Chinese auditory rehabilitation and CI programs will shine a light on the developmental pathway of healthcare infrastructure to meet emerging needs of the hearing-impaired population in other developing countries.

Mack I, Burckhardt MA, Heininger U, Prüfer F, Schulzke S, Wellmann S.

Symptomatic congenital cytomegalovirus infection in children of seropositive women.

Front Pediatr. 2017 Jun 9;5:134. doi: 10.3389/fped.2017.00134. eCollection 2017.

Cytomegalovirus (CMV) is the most frequent congenital virus infection worldwide. The risk of congenital CMV (cCMV) transmission is highest in seronegative women who acquire primary CMV infection during pregnancy. A growing body of evidence indicates that secondary CMV infections in pregnant women with preconceptual immunity (either through reactivation of latent virus or re-infection with a new strain of CMV) contribute to a much greater proportion of symptomatic cCMV than was previously thought. Here, we describe a case of symptomatic cCMV infection in the newborn of a woman with proven immunity prior to pregnancy. Diagnosis was confirmed by CMV PCR from amniotic fluid and fetal MR imaging. The newborn presented with typical cCMV symptoms including jaundice, hepatosplenomegaly, cholestasis, petechiae, small head circumference, and sensorineural hearing loss, the most common neurologic sequela. CMV was detected in infant blood and urine by PCR, and intravenous ganciclovir was initiated and continued orally for 6 weeks totally. Apart from persisting right-sided deafness, the child exhibited normal neurological development up through the last follow-up at 4.5 years. To date, the most effective strategy to prevent vertical CMV transmission is hygiene counseling for women of childbearing age, which, in our case, and in concordance with recent literature, applies to seronegative, as well as seropositive, women. Once an expecting mother shows seroconversion or signs of an active CMV infection, there are no established procedures to reduce the risk of transmission, or therapeutic options for the fetus with signs of infection. After birth, symptomatic infants can be treated with ganciclovir to inhibit viral replication and improve hearing ability and neurodevelopmental outcome. A comprehensive review of the literature, including our case study, reveals the most current and significant diagnostic and treatment options available. In conclusion, the triad of maternal hygiene counseling, postnatal hearing screening of all newborns, followed by CMV PCR in symptomatic infants, and antiviral therapy of infants with symptomatic cCMV provides an outline of best practice to reduce the burden of CMV transmission sequelae.

Martínez-Cruz CF, Poblano A, García-Alonso Themann P.

Changes in tonal audiometry in children with progressive sensorineural hearing loss and history of Neonatal Intensive Care Unit discharge. A 20 year long-term follow-up.

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:235-240. doi: 10.1016/j.ijporl.2017.08.022. Epub 2017 Aug 29.

OBJECTIVE: Newborns from Neonatal intensive care units (NICU) are at high-risk for sensorineural hearing loss (SNHL) a follow-up is needed for early diagnosis and intervention. Our objective here was to describe the features and changes of SNHL at different periods during a follow-up of almost 20 years.

METHODS: Risk factors for SNHL during development were analyzed. The audiological examination included: Brainstem auditory evoked potentials (BAEP), and Transient evoked otoacoustic emissions (TEOAE). At birth; tonal audiometry (between 125 and 8000 Hz), and tympanometry were performed at 5, 10, 15, and 20 years of age.

RESULTS: Sixty-five percent of cases presented bilateral absence of BAEP. At 5 years of age, the most frequent SNHL level was severe (42.5%), followed by moderate (22.5%), and profound level (20%), in all cases, the SNHL was symmetrical with a predominance of lesion for the high frequencies. Exchange transfusion was associated with a higher degree of SNHL (OR = 6.00, CI = 1.11-32.28, $p < 0.02$). In 55%, SNHL remained stable, but in 40% of the cases it was progressive. At the end of the study six cases with moderate loss progressed to the severe level and seven cases with severe level progressed to profound.

CONCLUSIONS: Forty percent of infants with SNHL discharged from NICU may present a progression in the hearing loss. Exchange transfusion was associated with a higher degree of SNHL. NICU graduates with SNHL merit a long-term audiological follow-up throughout their lifespan.

Matulat P, Stroe S, Am Zehnhoff-Dinnesen A.

[Transregional tracking in newborn hearing screening.](#)

HNO. 2017 Sep 22. doi: 10.1007/s00106-017-0424-y. [Epub ahead of print]

[Article in German]

BACKGROUND AND OBJECTIVE: When patients in the universal newborn hearing screening program move from one geographical area to another between initial screening and medical follow-up, the responsibility for their tracking also moves from one screening center to another. As a result, these patients are lost to follow-up according to the center which had initial responsibility. In cooperation with the Association of German Hearing Screening Centers ("Verband Deutscher Hörscreening-Zentralen e. V.," VDHZ) as an offer to the developers of tracking software, a concept for nationwide tracking including a reference implementation and evaluation is described.

METHODS: On the basis of error analysis of real screening data, techniques for preprocessing data, the technical background of the interface, and details regarding integration of the interface into tracking software are presented. Data from a stress test are shown.

RESULTS: In a simulation stress test with six hearing screening centers and 54,551 children, all requests were answered within an average response time of 637 ms (standard deviation, SD = 266 ms; median 613 ms). Anonymized surnames ($n = 675/1.24\%$) and duplicate entries in the database ($n = 49/0.01\%$) were detected.

CONCLUSION: A transregional tracking procedure using heterogeneous tracking software is possible without the use of a standardized screening ID. The presented approach seems conceptually and technically suitable.

Meyer L, Sharon B, Huang TC, Meyer AC, Gravel KE, Schimmenti LA, Swanson EC, Herd HE, Hernandez-Alvarado N, Coverstone KR, McCann M, Schleiss MR.

[Analysis of archived newborn dried blood spots \(DBS\) identifies congenital cytomegalovirus as a major cause of unexplained pediatric sensorineural hearing loss.](#)

Am J Otolaryngol. 2017 Sep - Oct;38(5):565-570. doi:10.1016/j.amjoto.2017.06.002. Epub 2017 Jun 7.

PURPOSE: Congenital cytomegalovirus (cCMV) infection is the most common non-genetic cause of sensorineural hearing loss (SNHL). However, accurate diagnosis of cCMV as the etiology of SNHL is problematic beyond the neonatal period. This study therefore examined whether cCMV infection could be identified retrospectively in children presenting with unexplained SNHL to a multidisciplinary diagnostic outpatient otolaryngology clinic at an academic medical center in Minnesota.

METHODS: Over a 4-year period, 57 patients with an age range of 3 months to 10 years with unexplained SNHL were recruited to participate in this study. Informed consent was obtained to test the archived dried blood spots (DBS) of these patients for cCMV infection by real-time PCR, targeting a highly conserved region of the CMV UL83 gene. **RESULTS** were normalized to recovery of an NRAS gene control. Chart review was conducted to identify subjects who underwent genetic testing and/or neurodiagnostic imaging to investigate possible genetic, syndromic, or anatomical causes of SNHL.

RESULTS: In total, 15 of the 57 children with unexplained SNHL tested positive for CMV DNA in their DBS (26%). A mean viral load of 8.3×10^4 ($\pm 4.1 \times 10^4$) [range, 1×10^3 - 6×10^5] copies/ μg DNA was observed in subjects retrospectively diagnosed with cCMV. No statistically significant correlation was found between viral load and SNHL severity.

CONCLUSIONS: A retrospective DBS analysis demonstrated that 26% of patients presenting with unexplained SNHL in childhood had cCMV. DBS testing is useful in the retrospective diagnosis of cCMV, and may provide definitive diagnostic information about the etiology of SNHL.

Müller J, Fechner H, Köhn A, Rißmann A.

Newborn hearing screening – Results of a parental survey in Saxony-Anhalt.

[Article in German]

Gesundheitswesen. 2017 May;79(5):388-393. doi: 10.1055/s-0035-1549969. Epub 2015 Jun 25.

BACKGROUND: In recent years quality assurance has become an essential part of today's health-care system in the wake of the modern patient-oriented quality management. With the statutory introduction of newborn hearing screening (NHS) in 2009, a quality assurance of these early detection methods has become necessary. The aim of the study was to determine patient satisfaction in relation to the NHS in Saxony-Anhalt.

PATIENTS/METHODS: During the period from November 2013 to April 2014, 394 parents were retrospectively interviewed about their experiences and expectations in relation to the NHS, using a standardised questionnaire. In total, 21 child care centres and 6 paediatric primary care centres from all over Saxony-Anhalt were involved.

RESULTS: It turns out that the majority of parents are satisfied with the NHS and 97.7% are in favour of the offer of an NHS. Of the surveyed parents, 69.3% felt the information as sufficient. However, only 66.2% of parents took a closer look at the leaflet issued by the G-BA. In addition, 17.7% of respondents are dissatisfied with the professional competence of the examining staff.

CONCLUSION: The study shows that the general attitude among parents towards newborn hearing screening was very positive. They felt reassured by it although there are some aspects still open to criticism.

Netten AP, Rieffe C, Ketelaar L, Soede W, Gadov 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. 2017 Oct 4. doi: 10.1097/AUD.0000000000000500. [Epub ahead of print]

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.

Pan J, Xu P, Tang W, Cui Z, Feng M, Wang C.

[Mutation analysis of common GJB2, SLC26A4 and 12S rRNA genes among 380 deafness patients in northern China.](#)

Int J Pediatr Otorhinolaryngol. 2017 Jul;98:39-42. doi: 10.1016/j.ijporl.2017.04.018. Epub 2017 Apr 12.

OBJECTIVES: The molecular etiology of nonsyndromic deafness in Chinese population has not been investigated systematically, our study is aim to investigate the molecular etiology of nonsyndromic deafness patients from Northern China (Heilongjiang province), in order to provide genetic test and counseling to families.

METHODS: 380 unrelated patients with hearing loss who attended to the Department of Otolaryngology, The Fourth Affiliated Hospital of Harbin Medical University were enrolled to our study. All patients were diagnosed with nonsyndromic deafness by audiologic evaluation, 202 normal-hearing individuals were taken as controls. Mutations in three common deafness-causing genes (GJB2, SLC26A4 and 12S rRNA) were screened by direct sequencing.

RESULTS: Mutations (homozygote or compound heterozygote) in GJB2 accounted for 8.9% (34/380) of the patients, mutations in SLC26A4 accounted for 10.0% (38/380) of the patients screened. Only one case was found to carry 12S rRNA 1555A > G (1/380, 0.26%). Five types of mutations in GJB2 were identified, GJB2 235delC was the most prevalent mutation in our patient group (76/380, 20.0%), followed by 299-300delAT with a frequency of 7.4% (28/380). Two types of mutations in SLC26A4 were detected in our patient group (IVS7-2A > G and 2168A > G). IVS7-2A > G was identified in 27 patients (27/380, 7.1%) and 2168A > G was identified in 14 patients (14/380, 3.7%).

CONCLUSIONS: Our results demonstrate that 19.2% patients with nonsyndromic deafness were caused by mutations in three common deafness genes (GJB2, SLC26A4 and 12S rRNA) in our northern China patient group. GJB2 235delC was the most prevalent mutation, same as in the most Asian populations. These data enrich the database of deafness mutations and provide the standard for clinical diagnose, treatment and genetic counseling in Northern China population.

Paul A, Prasad C, Kamath SS, Dalwai S, C Nair MK, Pagarkar W.

[Consensus statement of the Indian Academy of Pediatrics on Newborn Hearing Screening.](#)

Indian Pediatr. 2017 Aug 15;54(8):647-651. Epub 2017 Jun 4.

JUSTIFICATION: Hearing impairment is one of the most critical sensory impairments with significant social and psychological consequences.

Evidence-based, standardized national guidelines are needed for professionals to screen for hearing impairment during the neonatal period.

PROCESS: The meeting on formulation of national consensus guidelines on developmental disorders was organized by Indian Academy of Pediatrics in Mumbai, on 18th and 19th December, 2015. The invited experts included Pediatricians, Developmental Pediatricians, Pediatric Neurologists and Clinical Psychologists. The participants framed guidelines after extensive discussions.

OBJECTIVE: To provide guidelines on newborn hearing screening in India.

RECOMMENDATIONS: The first screening should be conducted before the neonate's discharge from the hospital - if it 'fails', then it should be repeated after four weeks, or at first immunization visit. If it 'fails' again, then Auditory Brainstem Response (ABR) audiometry should be conducted. All babies admitted to intensive care unit should be screened via ABR. All babies with abnormal ABR should undergo detailed evaluation, hearing aid fitting and auditory rehabilitation, before six months of age. The goal is to screen newborn babies before one month of age, diagnose hearing loss before three months of age and start intervention before six months of age.

Pimperton H, Kreppner J, Mahon M, Stevenson J, Terleksi E, Worsfold S, Yuen HM, Kennedy CR.

[Language outcomes in deaf or hard of hearing teenagers who are spoken language users: Effects of universal newborn hearing screening and early confirmation.](#)

Ear Hear. 2017 Sep/Oct;38(5):598-610. doi: 10.1097/AUD.0000000000000434.

OBJECTIVES: This study aimed to examine whether (a) exposure to universal newborn hearing screening (UNHS) and b) early confirmation of hearing loss were associated with benefits to expressive and receptive language outcomes in the teenage years for a cohort of spoken language users. It also aimed to determine whether either of these two variables was associated with benefits to relative language gain from middle childhood to adolescence within this cohort.

DESIGN: The participants were drawn from a prospective cohort study of a population sample of children with bilateral permanent childhood hearing loss, who varied in their exposure to UNHS and who had previously had their language skills assessed at 6-10 years. Sixty deaf or hard of hearing teenagers who were spoken language users and a comparison group of 38 teenagers with normal hearing completed standardized measures of their receptive and expressive language ability at 13-19 years.

RESULTS: Teenagers exposed to UNHS did not show significantly better expressive (adjusted mean difference, 0.40; 95% confidence interval [CI], -0.26 to 1.05; $d = 0.32$) or receptive (adjusted mean difference, 0.68; 95% CI, -0.56 to 1.93; $d = 0.28$) language skills than those who were not. Those who had their hearing loss confirmed by 9 months of age did not show significantly better expressive (adjusted mean difference, 0.43; 95% CI, -0.20 to 1.05; $d = 0.35$) or receptive (adjusted mean difference, 0.95; 95% CI, -0.22 to 2.11; $d = 0.42$) language skills than those who had it confirmed later. In all cases, effect sizes were of small size and in favor of those exposed to UNHS or confirmed by 9 months. Subgroup analysis indicated larger beneficial effects of early confirmation for those deaf or hard of hearing teenagers without cochlear implants ($N = 48$; 80% of the sample), and these benefits were significant in the case of receptive language outcomes (adjusted mean difference, 1.55; 95% CI, 0.38 to 2.71; $d = 0.78$). Exposure to UNHS did not account for significant unique variance in any of the three language scores at 13-19 years beyond that accounted for by existing

language scores at 6-10 years. Early confirmation accounted for significant unique variance in the expressive language information score at 13-19 years after adjusting for the corresponding score at 6-10 years (R change = 0.08, p = 0.03).

CONCLUSIONS: This study found that while adolescent language scores were higher for deaf or hard of hearing teenagers exposed to UNHS and those who had their hearing loss confirmed by 9 months, these group differences were not significant within the whole sample. There was some evidence of a beneficial effect of early confirmation of hearing loss on relative expressive language gain from childhood to adolescence. Further examination of the effect of these variables on adolescent language outcomes in other cohorts would be valuable.

Quevedo ALA, Leotti VB, Goulart BNG.

[Analysis of prevalence of self-reported hearing loss and associated factors: Primary versus proxy informant. \[Article in Portuguese\]](#)

Cad Saude Publica. 2017 Jul 3;33(6):e0076216. doi: 10.1590/0102-311X00076216.

The objective was to evaluate differences between prevalence rates for self-reported hearing loss and associated factors, obtained from responses by primary and proxy informants in a Population-Based Study on Human Communication Disorders (DCH-POP in Portuguese). This was a study on epidemiological methods using data from a cross-sectional household survey with a sample of 1,253 individuals from Southern Brazil. To verify differences between prevalence rates comparing primary informants and proxy informants, we used the chi-square or Fisher's exact test for categorical variables and Mann-Whitney for continuous variables. The log-binomial model was adjusted for hearing loss as the dependent variable, considering three datasets: the entire sample, only primary informants, and only proxy informants, estimating association by prevalence ratios. In the final models, only the independent variables age and dizziness were associated with hearing loss, independently of the dataset that was used. Proxy informants generally underestimated the prevalence rates for the target outcomes, when compared to primary informants.

Ricalde RR, Chiong CM, Labra PJP.

[Current assessment of newborn hearing screening protocols.](#)

Curr Opin Otolaryngol Head Neck Surg. 2017 Oct;25(5):370-377. doi:10.1097/MOO.0000000000000389.

PURPOSE OF REVIEW: The objective of this article is to assess current newborn hearing screening protocols. We will focus on technologies or modalities used, protocol steps, training of screeners, timing of first screen, and loss to follow-up. A summary of program reports focusing on protocols from Greece, China, South Africa, France, Spain, South Korea, Denmark, Italy, Turkey, Taiwan, South Korea, Poland and Iran as they are recently reported will also be presented.

RECENT FINDINGS: Community-based hearing screening programs in South Africa and efforts in the Asian region are being reported. The use of automated auditory brainstem response and staged procedures are gaining popularity because of low refer rates. However, follow-up issues remain a problem. The importance of having trained nonprofessional screeners and an efficient database is becoming more evident as the number of newborns screened for hearing loss increase each year.

SUMMARY: There are many reported protocols using different technologies, involving several stages, implemented in different settings which should not confuse but rather guide stakeholders so that programs may attain certain benchmarks and ultimately help the hard-of-hearing child in achieving his or her full potential.

Robertson VS, von Hapsburg D, Hay JS.

[The effect of hearing loss on novel word learning in infant- and adult-directed speech.](#)

Ear Hear. 2017 Jun 22. doi: 10.1097/AUD.0000000000000455. [Epub ahead of print]

OBJECTIVES: Relatively little is known about how young children with hearing impairment (HI) learn novel words in infant- and adult-directed speech (ADS). Infant-directed speech (IDS) supports word learning in typically developing infants relative to ADS. This study examined how children with normal hearing (NH) and children with HI learn novel words in IDS and ADS. It was predicted that IDS would support novel word learning in both groups of children. In addition, children with HI were expected to be less proficient word learners as compared with their NH peers.

DESIGN: A looking-while-listening paradigm was used to measure novel word learning in 16 children with sensorineural HI (age range 23.2 to 42.1 months) who wore either bilateral hearing aids (n = 10) or bilateral cochlear implants (n = 6) and 16 children with NH (age range 23.1 to 42.1 months) who were matched for gender, chronological age, and maternal education level. Two measures of word learning were assessed (accuracy and reaction time). Each child participated in two experiments approximately 1 week apart, one in IDS and one in ADS.

RESULTS: Both groups successfully learned the novel words in both speech type conditions, as evidenced by children looking at the correct picture significantly above chance. As a group, children with NH outperformed children with HI in the novel word learning task; however, there were no significant differences between performance on IDS versus ADS. More fine-grained time course analyses revealed that children with HI, and particularly children who use hearing aids, had more difficulty learning novel words in ADS, compared with children with NH.

CONCLUSIONS: The pattern of results observed in the children with HI suggests that they may need extended support from clinicians and caregivers, through the use of IDS, during novel word learning. Future research should continue to focus on understanding the factors (e.g., device type and use, age of intervention, audibility, acoustic characteristics of input, etc.) that may influence word learning in children with HI in both IDS and ADS.

Rütten H, Rissmann A, Brett B, Costa SD, Doßow B, Färber J, Fest S, Fritsch C, Lux A, Päge I, Spillner C, Redlich A.

[Congenital cytomegalovirus infection in Central Germany: An underestimated risk.](#)

Arch Gynecol Obstet. 2017 Aug;296(2):231-240. doi: 10.1007/s00404-017-4435-4. Epub 2017 Jun 17.

PURPOSE: This is the first study to determine the cytomegalovirus (CMV) seronegativity rate for women of childbearing age in Saxony-Anhalt and to determine the prevalence of clinically relevant congenital CMV (cCMV) infection in Central Germany, because there are no valid data available.

METHODS: The retrospective study was undertaken between January 2005 and December 2015. For the first time in Germany, the following seven data sources were used to analyze the prevalence of clinically relevant cCMV infection and the rate of CMV seronegative women of childbearing age: CMV Screening in maternity unit, University Women's Hospital, Social Paediatrics Centre (SPC), Malformation Monitoring Centre (MMC), Newborn Hearing Screening (NHS), Neonatal Intensive Care Unit (NICU), and In-house Doctor Department. Key parameters were anti-CMV IgG and IgM, CMV PCR of urine, and clinically relevant symptoms caused by CMV.

RESULTS: Between 46 and 52% of women of childbearing age were CMV seronegative. The prevalence of clinically relevant cCMV infection was between 0.008 and 0.04%.

CONCLUSIONS: The CMV seronegativity rate of women of childbearing age was confirmed to be in the middle range of estimated data from other sources in Germany. Data from the NICU, SPC, NHS, and MMC show the prevalence of clinically relevant cCMV infection. The risk of all cCMV

infections is underestimated. Thus, the true prevalence of clinically relevant and subclinical cCMV infections is >0.04%.

Sabbag JC, Lacerda ABM.

[Neonatal hearing screening in primary health care and family health care. \[Article in English, Portuguese\]](#)

Codas. 2017 Aug 10;29(4):e20160102. doi: 10.1590/2317-1782/20172016102.

PURPOSE: The Universal Newborn Hearing Screening (UNHS) looks for early diagnosis and rehabilitation of newborns at risk or not of hearing impairment. The purpose is analyze the flow of Universal Newborn Hearing Screening in the family health care strategy unit through the tracking and monitoring of children.

METHODS: This is a quantitative and retrospective study. The trace begins with the third copy of the Live Newborn Declaration, filled in at the maternity ward. An interview with parents and guardians was made by a community agent at the Health Unit or at the home of the newborn. Monitoring was conducted by live birth declaration and the information collected by the interviewer from maternal and child health booklet and the follow-up at high complexity services.

RESULTS: The sample was made up of 50 neonates. 52% were between 30 and 89 days and 54% were male. 12% of newborns presented a risk factor for hearing loss and the neonatal screening was performed in 86% of cases. Hearing health measures show integrality in hearing impairment care at the basic health unit to high complexity hospital.

CONCLUSION: The flow of care for newborn hearing screening is in agreement with the child health care guidelines in Curitiba, however, it is not yet universal. In conclusion, the participation of the family health strategy unit in the tracking and monitoring of children submitted to the Universal newborn hearing screening program is feasible and recommended.

Sachdeva K, Sao T.

[Outcomes of newborn hearing screening program: A hospital based study.](#)

Indian J Otolaryngol Head Neck Surg. 2017 Jun;69(2):194-198. doi: 10.1007/s12070-017-1062-0. Epub 2017 Jan 17.

Hearing loss is hidden disability and second most common congenital pathology. Prevention, early identification and early intervention of hearing loss can prevent further disability in development of speech, language, cognition and other developmental domains. The prevalence of congenital hearing loss has been estimated to be 1.2-5.7 per thousand in neonates. In these contexts, the aim of study was to determine outcomes of neonates hearing screening program in Hospital. It is a clinical cross-sectional study which was conducted in tertiary care centre from 8th July, 2015 to 31st May, 2016. Total no of 2254 cases were screened. Details case history including high risk register, Pediatric Audiometry, Otoacoustic Emission tests were performed followed brainstem evoked response audiometry. The Prevalence of hearing loss among high risk babies confirmation by BERA was 8.8% per 1000 babies and 16 cases were recommended for Cochlear Implant. The screening protocol with objective test i.e. Distortion Product Otoacoustic Emission and confirmation by Brainstem Evoked Response Audiometry is very useful tool in early identification of congenital hearing loss in neonates. Hence, the results of this study will be used to initiate universal newborn hearing screening in other hospitals. Moreover, this study highlights the relevance of neonatal hearing screening in other states of India and country where this screening is not performed routinely in all hospitals and creating awareness to identify neonatal risk factors associated with hearing loss and understand the importance of early identification and early intervention and among health care professionals.

Saki N, Bayat A, Hoseinabadi R, Nikakhlagh S, Karimi M, Dashti R.

[Universal newborn hearing screening in southwestern Iran.](#)

Int J Pediatr Otorhinolaryngol. 2017 Jun;97:89-92. doi: 10.1016/j.ijporl.2017.03.038. Epub 2017 Mar 31.

OBJECTIVES: The implementation of Neonatal Hearing Screening (NHS) program is still at the preliminary stage particularly in developing countries despite the burden of permanent congenital and early-onset hearing impairment. The purpose of this study was to report results for universal newborn NHS in a cohort of children born in the southwestern region of Iran, as part of a national screening program set up by the Iranian National Health System.

METHODS: During this cross-sectional study, which took place between March 2013 and April 2016, healthy newborns were screened using transient evoked otoacoustic emissions (TEOAEs) and automated auditory brainstem responses (AABRs) methods at several points in time as early as possible after birth. Screening followed a two-stage strategy and newborns referred after the second-stage screening were scheduled for diagnostic evaluation.

RESULTS: A total of 92,521 newborns were screened in the urban (n = 67,780) and rural (n = 24,741) regions. Hearing impairment was confirmed in 223 (2.41 per 1000) newborns. One hundred forty-one (1.52 per 1000) of these newborns were affected bilaterally. More than 87% of these infants (195/223) showed a sensorineural hearing loss, while the defect was found to be conductive in 12 cases (P < 0.001). Of the 223 cases with hearing loss, 28 (12.5%) infants had auditory neuropathy. The majority of the infants, in both urban and rural regions, showed severe hearing impairment. We did not observe any significant difference among the incidences associated with gender (p = 0.29).

CONCLUSION: Our results demonstrated that universal newborn hearing screening program is an adequate program for southwestern of Iran with high coverage, low referral rate, and good follow-up rate.

Sanyelbhaa H, Kabel A, Abo El-Naga HAE, Sanyelbhaa A, Salem H.

[The risk ratio for development of hereditary sensorineural hearing loss in consanguineous marriage offspring.](#)

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:7-10. doi: 10.1016/j.ijporl.2017.07.020. Epub 2017 Jul 19.

OBJECTIVES: This study aims to define the relative risk of development of hearing loss in offspring of consanguineous marriages.

MATERIALS AND METHODS: This is a retrospective case-control study conducted in a tertiary referral center in Jeddah, KSA. The study group included 1600 probands (848 males, 752 females), with age range 0.5-12 years (6.6 ± 3.6). The study group comprised of two equal, age and sex matched subgroups; Hearing Loss (HL) group and Normal Hearing (NH) group. The children included in the HL group should have idiopathic or non syndromic genetic sensorineural hearing loss.

RESULTS: The HL Group comprised 800 children with variable degrees of sensorineural hearing loss. Profound and severe degrees of hearing loss were the most prevalent degrees (P < 0.05%). The prevalence of consanguineous marriage offspring in the NH group was 42.5%, while in the HL group it was 68.9% (P < 0.05). The differences between both study subgroups regarding the distribution of different degrees of parental consanguinity (first, second, double first, and first once removed cousins) were insignificant (P > 0.05). The relative risk and 95% confidence interval (RR, 95% CI) for development of hearing loss in offspring of consanguineous marriage was 1.76 (95% CI 1.57-1.97, P < 0.001).

CONCLUSIONS: There was 76% increased risk for consanguineous marriage progeny to develop SNHL when compared to non consanguineous progeny.

Serra A, Spinato G, Cocuzza S, Licciardello L, Pavone P, Maiolino L.

[Adaptive psychological structure in childhood hearing impairment: audiological correlations.](#)

Acta Otorhinolaryngol Ital. 2017 Jun;37(3):175-179. doi: 10.14639/0392-100X-1291.

The present research deals with the clinical and social problems present during linguistic and cognitive development of deaf children. Currently, the development of Theory of Mind represents an important research field in deafness studies. These international studies highlighted a significant alteration in the development of Theory of Mind in deaf children compared to normal hearing children, especially in cases of congenital or preverbal hearing loss. In particular, the research focuses on the skills of deaf children in recognising emotions and desires, through both perceptive and cognitive methods, by evaluation of psycho-cognitive skills of children with severe hearing loss using a set of questions to be administered to hearing loss patients. The experiment was performed on a group composed of 10 children (5 males and 5 females) aged 4 to 9 years and 54 to 108 months, affected by bilateral congenital hearing loss (severe to total), or hearing loss that developed in preverbal children the year before entering elementary school, or during the fourth year of elementary school. The selection criteria were based on: audiologic evaluation, neuro-psychological tests administered to assess general, cognitive as well as praxis and perceptive abilities, and clinical observations performed to assess psychopathology using tests that assess development of both visual perceptive (Coloured Progressive Matrices) and graphic representational abilities (Test of Human Figure Drawings and the Family Drawing Test). The instrument "cognitive" was the "Deaf Children Series", arranged by us, that consists of a mental status examination (MSE) that evaluates: level of cognitive (knowledge-related) ability, emotional mood, and speech and thought patterns at the time of evaluation. Deaf children show a reduced responsiveness to the expressions of sadness on the perceptive side. Through the test, we observed a psychodynamic defense mechanism considering perceptive understanding performance. On the contrary, in normal hearing children, the emotion 'fear' is the most difficult to identify. Deaf children seem to be more susceptible to recognition of visual emotions. Furthermore, deaf children present significant problem-solving skills and emotional recognition skills, possibly as a result of their hearing impairment.

Su BM, Park JS, Chan DK.

[Impact of primary language and insurance on pediatric hearing health care in a multidisciplinary clinic.](#)

Otolaryngol Head Neck Surg. 2017 Oct;157(4):722-730. doi: 10.1177/0194599817725695. Epub 2017 Sep 12.

OBJECTIVE: This study aims to describe the effects of primary language and insurance status on care utilization among deaf or hard-of-hearing children under active otolaryngologic and audiologic care. Study Design Cross-sectional analysis. Setting Multidisciplinary hearing loss clinic at a tertiary center.

SUBJECTS AND METHODS: Demographics, hearing loss data, and validated survey responses were collected from 206 patients aged 0 to 19 years. Two-sided t tests and χ^2 tests were used to obtain descriptive statistics and hypothesis testing.

RESULTS: Of the sample, 52.4% spoke primarily English at home. Non-English-speaking children and families were less likely to receive psychiatric counseling (12.2% vs 35.2% in the English group, $P < .001$) and reported more difficulty obtaining educational interventions ($P = .016$), and 68.9% had public insurance. Parents of publicly insured children were less likely to know the type or degree of their child's hearing loss (56.9% vs 75.4%, $P = .022$), and these children were older on presentation to the clinic (8.5 vs 6.5 years of age, $P = .01$) compared to privately insured children. Publicly insured children were less likely to receive cochlear implants ($P = .046$) and reported increased difficulty obtaining hearing aids ($P = .047$). While all patients reported impairment in hearing-related quality of life, publicly insured children aged 2 to 7 years were more likely to perform below minimum thresholds on measures of auditory/oral functioning.

CONCLUSION: Even when under active care, deaf or hard-of-hearing children from families who do not speak English at home or with public insurance face more difficulty obtaining educational services, cochlear implants, and hearing aids. These findings represent significant disparities in access to necessary interventions.

van Beeck Calkoen EA, Sanchez Aliaga E, Merkus P, Smit CF, van de Kamp JM, Mulder MF, Goverts ST, Hensen EF.

[High prevalence of abnormalities on CT and MR imaging in children with unilateral sensorineural hearing loss irrespective of age or degree of hearing loss.](#)

Int J Pediatr Otorhinolaryngol. 2017 Jun;97:185-191. doi:10.1016/j.ijporl.2017.04.002. Epub 2017 Apr 4.

OBJECTIVE: Evaluation of causal abnormalities identified on CT and MR imaging in children with unilateral sensorineural hearing loss (USNHL), and the association with age and severity of hearing loss.

STUDY DESIGN: Retrospective cohort study.

SETTING: Tertiary referral otology/audiology center.

PATIENTS AND DIAGNOSTIC INTERVENTIONS: 102 children diagnosed with USNHL between 2006 and 2016 were included. They underwent CT and/or MR imaging for the evaluation of the etiology of their hearing loss.

MAIN OUTCOME MEASURES: Radiologic abnormalities of the inner ear and brain associated with USNHL.

RESULTS: Using CT and/or MR imaging, causal abnormalities were identified in 49%, which is higher than previously reported (25-40%). The most frequently affected site was the labyrinth (29%), followed by the cochlear nerve (9%) and brain (7%). No significant difference in the number or type of abnormalities was found for the degree of hearing loss or age categories.

CONCLUSIONS: Imaging is essential in the etiologic analysis of USNHL because of the high prevalence of causative abnormalities that can be identified with radiology, irrespective of the patients' age or degree of hearing loss. CT and MR imaging are complementary imaging options. The ideal imaging algorithm is controversial. Based on our findings, we conclude that there is limited additional diagnostic value of simultaneous dual modality imaging over sequential diagnostics. We therefore perform a stepwise radiological workup in order to maximize the diagnostic yield while minimizing impact and costs. If the primary imaging modality does not identify a cause for USNHL, performing the alternative imaging modality should be considered.

van Noort-van der Spek IL, Goedegebure A, Hartwig NG, Kornelisse RF, Franken MJP, Weisglas-Kuperus N.

[Normal neonatal hearing screening did not preclude sensorineural hearing loss in two-year-old very preterm infants.](#)

Acta Paediatr. 2017 Oct;106(10):1569-1575. doi: 10.1111/apa.13960. Epub 2017 Jul 13.

AIM: Very preterm infants are at risk of neonatal hearing loss. However, it is unknown whether infants with a normal neonatal hearing screening result risk sensorineural hearing loss (SNHL) at a later age.

METHODS: This cohort study was conducted at the Erasmus Medical University Center Rotterdam, the Netherlands, on 77 very preterm infants born between October 2005 and September 2008. All infants underwent auditory brainstem response audiometry during neonatal hearing screening and at two years of corrected age. The frequency of SNHL in infants with a normal neonatal hearing screening was analysed and the risk factors associated with newly diagnosed SNHL in these infants were examined.

RESULTS: We found that 3.9% (3/77) of the very preterm infants showed permanent hearing loss during their neonatal hearing screening. In addition, a relatively high prevalence of newly diagnosed SNHL (4.3%) was found in three of the 70 infants followed up at the age of two. The total prevalence rate of permanent hearing loss in the cohort was approximately 8%.

CONCLUSION: A normal outcome of neonatal hearing screening did not guarantee normal hearing at two years of age in this very preterm cohort and paediatricians should be alert to the possibility of late-onset SNHL.

Vila PM, Ghogomu NT, Odom-John AR, Hullar TE, Hirose K.

Infectious complications of pediatric cochlear implants are highly influenced by otitis media.

Int J Pediatr Otorhinolaryngol. 2017 Jun;97:76-82. doi: 10.1016/j.ijporl.2017.02.026. Epub 2017 Mar 12.

OBJECTIVE: Determine the incidence of ear infections in cochlear implant patients, evaluate the contribution of otitis media to complications, describe the bacteriology of otitis media in the cochlear implant population, the treatment provided at our center, and the long term outcome.

METHODS: Data collected included age at implantation, history of otitis media or ear tubes, etiology of hearing loss, inner ear anatomy, postoperative infections, time to infection, route of antibiotic administration, and interventions for infections. Categories of infection were acute otitis media, otitis media with effusion, tube otorrhea, meningitis, scalp cellulitis, and infection at the implant site.

RESULTS: Middle ear infections were diagnosed in 37% of implanted ears. Extension of middle ear infections into the implant site occurred in 2.8% of all implants (n = 16). Of the 16 infected devices, 10 were successfully treated with antibiotic therapy and did not require explantation. The retained implant group and explanted group both included some middle ear microbes such as Haemophilus influenzae and Streptococcus pneumoniae, as well as skin flora such as Staphylococcus aureus.

CONCLUSION: Otitis media in pediatric cochlear implant patients is a common event and usually does not lead to complications of the cochlear implant. However, when the ear infection spreads to the scalp and the implant site, it is still possible to eliminate the infection using antibiotic therapy, particularly when treatment is directed to the specific organism that is recovered from the infected space and the duration and route of antibiotic treatment is carefully considered.

Välilmaa T, Kunnari S, Laukkanen-Nevala P, Lonka E; National Clinical Research Team.

Early vocabulary development in children with bilateral cochlear implants.

Int J Lang Commun Disord. 2017 Jun 16. doi: 10.1111/1460-6984.12322. [Epub ahead of print]

BACKGROUND: Children with unilateral cochlear implants (CIs) may have delayed vocabulary development for an extended period after implantation. Bilateral cochlear implantation is reported to be associated with improved sound localization and enhanced speech perception in noise. This study proposed that bilateral implantation might also promote early vocabulary development. Knowledge regarding vocabulary growth and composition in children with bilateral CIs and factors associated with it may lead to improvements in the content of early speech and language intervention and family counselling.

AIMS: To analyse the growth of early vocabulary and its composition during the first year after CI activation and to investigate factors associated with vocabulary growth.

METHODS & PROCEDURES: The participants were 20 children with bilateral CIs (12 boys; eight girls; mean age at CI activation = 12.9 months). Vocabulary size was assessed with the Finnish version of the MacArthur Communicative Development Inventories (CDI) Infant Form and compared with normative data. Vocabulary composition was analysed in relation to vocabulary size. Growth curve modelling was implemented using a linear mixed model to analyse the effects of the following variables on early vocabulary growth: time, gender, maternal education, residual hearing with hearing aids, age at first hearing aid fitting and age at CI activation.

OUTCOMES & RESULTS: Despite clear vocabulary growth over time, children with bilateral CIs lagged behind their age norms in receptive vocabulary during the first 12 months after CI activation. In expressive vocabulary, 35% of the children were able to catch up with their age norms, but 55% of the children lagged behind them. In receptive and expressive vocabularies of 1-20 words, analysis of different semantic categories indicated that social terms constituted the highest proportion. Nouns constituted the highest proportion in vocabularies of 101-400 words. The proportion of verbs remained below 20% and the proportion of function words and adjectives remained below 10% in the vocabularies of 1-400 words. There was a significant main effect of time, gender, maternal education and residual hearing with hearing aids before implantation on early receptive vocabulary growth. Time and residual hearing with hearing aids had a significant main effect also on expressive vocabulary growth.

CONCLUSIONS & IMPLICATIONS: Vocabulary development of children with bilateral CIs may be delayed. Thus, early vocabulary development needs to be assessed carefully in order to provide children and families with timely and targeted early intervention for vocabulary acquisition.

Wang L, Wang Z, Gao F, Peng KA.

Modiolar ossification in paediatric patients with auditory neuropathy.

J Laryngol Otol. 2017 Jul;131(7):598-601. doi: 10.1017/S0022215117001037. Epub 2017 May 15.

OBJECTIVE: To describe our finding of increased ossification of the modiolus in paediatric patients with auditory neuropathy who met criteria for cochlear implantation.

METHODS: A retrospective case series with a comparison group at a tertiary referral centre is described. Seven paediatric patients with auditory neuropathy who met criteria for and underwent cochlear implantation were identified. Fifteen paediatric implantees with bilateral profound sensorineural hearing loss were included as the comparison group. All patients underwent pre-operative computed tomography. Attenuation at the modiolus was measured in all subjects by a neuroradiologist blinded to clinical information.

RESULTS: Attenuation values in the modiolus in the auditory neuropathy patients (mean \pm standard deviation = 796.2 \pm 53.0 HU) was statistically significantly higher than in the comparison sensorineural hearing loss patients (267.1 \pm 45.6 HU; $p < 0.05$, t-test).

CONCLUSION: Patients with auditory neuropathy who meet criteria for cochlear implantation demonstrate significantly higher modiolar attenuation on computed tomography imaging, consistent with increased ossification at the modiolus.

Wang X, Wu D, Zhao Y, Li D, He D.

Knowledge and attitude of mothers regarding infant hearing loss in Changsha, Hunan province, China.

Int J Audiol. 2017 Oct 16:1-6. doi: 10.1080/14992027.2017.1366671. [Epub ahead of print]

OBJECTIVE: The objective of this study was to explore the knowledge and attitude among mothers of newborns regarding infant hearing loss (HL) in

Changsha, Hunan province, China.

DESIGN: A questionnaire including 18 items was given to mothers.

STUDY SAMPLE: A total of 115 mothers participated in the study.

RESULTS: Seven risk factors for hearing loss were identified correctly by above 60% of respondents and the top three were prolonged noise (88.7%), high fever (82.6%) and ear discharge (82.6%). Poor knowledge was demonstrated on risk factors jaundice (20.0%), measles (22.6%), convulsion (33.0%) and traditional Chinese medicine (39.1%). Maternal knowledge scores in identification and intervention (2.68 ± 0.31) was slightly higher than the score in risk factors (2.47 ± 0.34). Ninety-nine per cent of the mothers expressed the willingness to test baby's hearing soon after birth and concern about hearing.

CONCLUSIONS: Mothers were concerned about baby's hearing and the attitude was positive. However, the correct recognition rate towards some risk factors for HL was low. Action needs to be taken to raise awareness about ear and hearing care, prevent HL caused by preventable causes and prompt early identification, early diagnosis and intervention of HL.

Wang XY, Huang LH, Du YT.

[Research progress on the etiology of delayed-onset hearing loss in children.](#)

[\[Article in Chinese\]](#)

Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2017 Oct 7;52(10):787-791. doi: 10.3760/cma.j.issn.1673-0860.2017.10.017.

Newborn hearing screening is an effective method for early detection of hearing loss, however, it is not able to detect delayed-onset hearing loss. By exploring the etiology of delayed-onset hearing loss in children, it can provide a clinical basis for early detection of delayed-onset hearing loss. Mutations in SLC26A4, mitochondrial, GJB2 and other genes, enlarged vestibular aqueduct, congenital cytomegalovirus infection, extracorporeal membrane oxygenation, and auditory neuropathy et al were more commonly reported risk factors. In this paper, the risk factors related to delayed-onset hearing loss, which are divided into 5 categories: genetic mutation, abnormal inner ear malformation, perinatal factors, auditory neuropathy and no identifiable cause, are reviewed and analyzed.

Wien MA, Whitehead MT.

[The association among prematurity, cochlear hyperintensity, and hearing loss.](#)

Neuroradiol J. 2017 Oct;30(5):448-453. doi: 10.1177/1971400917709623. Epub 2017 Jun 20.

BACKGROUND Prematurity is a major risk factor for neonatal hearing loss. Recent advancements in magnetic resonance imaging (MRI) have made it possible to evaluate structural details of the membranous labyrinths in premature infants that have heretofore been inaccessible.

OBJECTIVE We compared the prevalence of abnormal cochlear signal intensity in premature and term neonates and evaluated for a potential link with hearing loss.

MATERIALS AND METHODS We retrospectively reviewed 148 consecutive MR exams performed in premature (< 37 weeks' gestation) and term neonates performed over a 30-month period. Cochlear signal alteration was evaluated on three-dimensional T1-weighted imaging (T1WI) sequences. Each patient's electronic medical record was reviewed to document demographics, symptomatology, physical exam findings, and potential medical variables that could contribute to cochlear signal alteration.

RESULTS Cochlear hyperintensity on T1WI was present in 6.8% patients (n = 10) overall, but was much more common in preterm than term patients (12.2% (9 of 74) vs. 1.4% (1 of 74), respectively; p value < 0.05; Fisher's exact test). Overall, 14.9% (n = 15) of the patients with hearing test results failed the screening Auditory Brainstem Response exam. However, failure was much more common among patients with cochlear hyperintensity on T1WI than those with normal findings (56% (5 of 9) vs. 11% (10 of 92), respectively; p value < 0.01; Fisher's exact test).

CONCLUSION Cochlear hyperintensity on T1WI is more common in preterm than term neonates, and potentially associated with hearing loss. Cochleae should be closely scrutinized in all premature infants; signal alterations should prompt further diagnostic inquiry and possible early otolaryngology referral.

Wong LY, Espinoza F, Alvarez KM, Molter D, Saunders JE.

[Otoacoustic emissions in rural Nicaragua: Cost analysis and implications for newborn hearing screening.](#)

Otolaryngol Head Neck Surg. 2017 May;156(5):877-885. doi:10.1177/0194599817696306. Epub 2017 Feb 1.

OBJECTIVE (1) Determine the incidence and risk factors for congenital hearing loss. (2) Perform cost analysis of screening programs. Study Design Proportionally distributed cross-sectional survey. Setting Jinotega, Nicaragua.

SUBJECTS & METHODS Otoacoustic emissions (OAEs) were used to screen 640 infants <6 months of age from neonatal intensive care unit, institutional, and home birth settings. Data on 15 risk factors were analyzed. Cost of 4 implementation strategies was studied: universal screening, screening at the regional health center (RHC), targeted screening, and screening at the RHC plus targeted screening. Cost-effectiveness analysis over 10 years was based on disability-adjusted life year estimates, with the World Health Organization standard of cost-effectiveness ratio (CER) / gross domestic product (GDP) < 3, with GDP set at \$4884.15.

RESULTS Thirty-eight infants failed the initial OAE (5.94%). In terms of births, 325 (50.8%) were in the RHC, 69 (10.8%) in the neonatal intensive care unit, and 29 (4.5%) at home. Family history and birth defect were significant in univariate analysis; birth defect was significant in multivariate analysis. Cost-effectiveness analysis demonstrated that OAE screening is cost-effective without treatment (CER/GDP = 0.06-2.00) and with treatment (CER/GDP = 0.58-2.52).

CONCLUSIONS Our rate of OAE failures was comparable to those of developed countries and lower than hearing loss rates noted among Nicaraguan schoolchildren, suggesting acquired or progressive etiology in the latter. Birth defects and familial hearing loss correlated with OAE failure. OAE screening of infants is feasible and cost-effective in rural Nicaragua, although highly influenced by estimated hearing loss severity in identified infants and the high travel costs incurred in a targeted screening strategy.

Wroblewska-Seniuk K, Greczka G, Dabrowski P, Szyfter-Harris J, Mazela J.

[Hearing impairment in premature newborns: Analysis based on the national hearing screening database in Poland.](#)

PLoS One. 2017 Sep 14;12(9):e0184359. doi: 10.1371/journal.pone.0184359. eCollection 2017.

OBJECTIVES: The incidence of sensorineural hearing loss is between 1 and 3 per 1000 in healthy neonates and 2-4 per 100 in high-risk infants.

The national universal neonatal hearing screening carried out in Poland since 2002 enables selection of infants with suspicion and/or risk factors of hearing loss. In this study, we assessed the incidence and risk factors of hearing impairment in infants ≤ 33 weeks' gestational age (wga).

METHODS: We analyzed the database of the Polish Universal Newborns Hearing Screening Program from 2010 to 2013. The study group involved 11438 infants born before 33 wga, the control group-1487730 infants. Screening was performed by means of transient evoked otoacoustic emissions. The risk factors of hearing loss were recorded. Infants who failed the screening test and/or had risk factors were referred for further audiological evaluation.

RESULTS: Hearing deficit was diagnosed in 11% of infants ≤ 25 wga, 5% at 26-27 wga, 3.46% at 28 wga and 2-3% at 29-32 wga. In the control group the incidence of hearing deficit was 0.2% (2.87% with risk factors). The most important risk factors were craniofacial malformations, very low birth weight, low Apgar score and mechanical ventilation. Hearing screening was positive in 22.42% newborns ≤ 28 wga and 10% at 29-32 wga and in the control group.

CONCLUSIONS: Hearing impairment is a severe consequence of prematurity. Its prevalence is inversely related to the maturity of the baby. Premature infants have many concomitant risk factors which influence the occurrence of hearing deficit.

Yoshinaga-Itano C, Sedey AL, Wiggin M, Chung W.

[Early hearing detection and vocabulary of children with hearing loss.](#)

Pediatrics. 2017 Aug;140(2). pii: e20162964. doi: 10.1542/peds.2016-2964. Epub 2017 Jul 8.

BACKGROUND AND OBJECTIVES: To date, no studies have examined vocabulary outcomes of children meeting all 3 components of the Early Hearing Detection and Intervention (EHDI) guidelines (hearing screening by 1 month, diagnosis of hearing loss by 3 months, and intervention by 6 months of age). The primary purpose of the current study was to examine the impact of the current EHDI 1-3-6 policy on vocabulary outcomes across a wide geographic area. A secondary goal was to confirm the impact of other demographic variables previously reported to be related to language outcomes.

METHODS: This was a cross-sectional study of 448 children with bilateral hearing loss between 8 and 39 months of age (mean = 25.3 months, SD = 7.5 months). The children lived in 12 different states and were participating in the National Early Childhood Assessment Project.

RESULTS: The combination of 6 factors in a regression analysis accounted for 41% of the variance in vocabulary outcomes. Vocabulary quotients were significantly higher for children who met the EHDI guidelines, were younger, had no additional disabilities, had mild to moderate hearing loss, had parents who were deaf or hard of hearing, and had mothers with higher levels of education.

CONCLUSIONS: Vocabulary learning may be enhanced with system improvements that increase the number of children meeting the current early identification and intervention guidelines. In addition, intervention efforts need to focus on preventing widening delays with chronological age, assisting mothers with lower levels of education, and incorporating adults who are deaf/hard-of-hearing in the intervention process.

Yuan EF, Xia W, Huang JT, Hu L, Liao X, Dai X, Liu SM.

[A sensitive and convenient method for clinical detection of non-syndromic hearing loss-associated common mutations.](#)

Gene. 2017 Sep 10;628:322-328. doi: 10.1016/j.gene.2017.07.045. Epub 2017 Jul 19.

BACKGROUND: The majority of non-syndromic hearing loss (NSHL) patients result from causative mutations in GJB2, SLC26A4 and mitochondrial 12S rRNA genes. Accurate detection of these genetic mutations is increasingly recognized for its clinical significance to reduce incidence and guide individual treatment of NSHL. Current methods for clinical practice are labor intensive, expensive or of low sensitivity.

METHODS: Genomic DNA from 7 newborns not passing the hearing screening and 94 newborns passing the hearing screening were analyzed for the common mutations using high resolution melting analysis (HRMA) and Sanger sequencing.

RESULTS: Our newly developed HRMA allowed the hot-spot mutations of GJB2 c.176_191del16 and c.235delC, SLC26A4 IVS7-2A>G and mitochondrial 12S rRNA 1494C>T and 1555A>G to be detected by melting profiles based on small amplicons. HRMA can distinguish different content mutant DNA from wildtype DNA, with a detection limit of 5%. Moreover, the results were highly concordant between HRMA and Sanger sequencing.

CONCLUSIONS: These results indicate that HRMA could be used as a routine clinical method for prenatal diagnosis and newborn genetic screening due to its accuracy, sensitivity, and rapid, low-cost and less laborious workflows.