

## What Are Others Publishing About Early Hearing Detection and Intervention?

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

Whereas JEHDI is the only journal that focuses specifically on improving EHD systems, many other journals publish relevant articles as a part their journal's broader focus. To help JEHDI readers stay up-to-date about recently published material, we provide titles and abstracts of what has been published in the last 9 months that JEHDI editors think are most relevant to improving EHD programs. Titles of all articles are hyperlinked to the source.

[Abdollahi FZ](#), Ahmadi T, [Manchaiah V](#), [Lotfi Y](#).

### [Auditory Brainstem Response Improvements in Hyperbilirubinemic Infants.](#)

*J Audiol Otol.* 2016 Apr;20(1):13-6. doi: 10.7874/jao.2016.20.1.13. Epub 2016 Apr 21.

**Background and Objectives:** Hyperbilirubinemia in infants have been associated with neuronal damage including in the auditory system. Some researchers have suggested that the bilirubin-induced auditory neuronal damages may be temporary and reversible. This study was aimed at investigating the auditory neuropathy and reversibility of auditory abnormalities in hyperbilirubinemic infants.

**Subjects and Methods:** The study participants included 41 full term hyperbilirubinemic infants (mean age 39.24 days) with normal birth weight (3,200-3,700 grams) that admitted in hospital for hyperbilirubinemia and 39 normal infants (mean age 35.54 days) without any hyperbilirubinemia or other hearing loss risk factors for ruling out maturational changes. All infants in hyperbilirubinemic group had serum bilirubin level more than 20 milligram per deciliter and undergone one blood exchange transfusion. Hearing evaluation for each infant was conducted twice: the first one after hyperbilirubinemia treatment and before leaving hospital and the second one three months after the first hearing evaluation. Hearing evaluations included transient evoked otoacoustic emission (TEOAE) screening and auditory brainstem response (ABR) threshold tracing.

**Results:** The TEOAE and ABR results of control group and TEOAE results of the hyperbilirubinemic group did not change significantly from the first to the second evaluation. However, the ABR results of the hyperbilirubinemic group improved significantly from the first to the second assessment ( $p=0.025$ ).

**Conclusion:** The results suggest that the bilirubin induced auditory neuronal damage can be reversible over time so we suggest that infants with hyperbilirubinemia who fail the first hearing tests should be reevaluated after 3 months of treatment.

Barreira-Nielsen C, Fitzpatrick E, Hashem S, Whittingham J, Barrowman N, Aglipay M.

### [Progressive Hearing Loss in Early Childhood.](#)

*Ear Hear.* 2016 Sep-Oct;37(5):e311-21. doi: 10.1097/AUD.0000000000000325.

**Objectives:** Deterioration in hearing thresholds in children is of concern due to the effect on language development. Before universal newborn hearing screening (UNHS), accurate information on the progression of hearing loss was difficult to obtain due to limited information on hearing loss onset. The objective of this population-based study was to document the proportion of children who experienced progressive loss in a cohort followed through a UNHS program in one region of Canada. We explored risk factors for progression including risk indicators, audiologic, and clinical characteristics of children. We also investigated deterioration in hearing as a function of age. For this study, two working definitions of progressive hearing loss were adopted: (1) a change of  $\geq 20$  dB in the 3 frequencies (500, 1000, and 2000 Hz) pure-tone average, and (2) a decrease of  $\geq 10$  dB at two or more adjacent frequencies between 500 and 4000 Hz or a decrease in 15 dB at one octave frequency in the same frequency range.

**Design:** Population-based data were collected prospectively on a cohort of children identified from 2003 to 2013 after the implementation of UNHS. Clinical characteristics including risk indicators (as per Joint Committee on Infant Hearing), age at diagnosis, type and severity of hearing loss, and initial audiologic information were recorded when children were first identified with hearing loss. Serial audiometric results were extracted from the medical charts for this study. Differences between children with progressive and stable hearing loss were explored using  $\chi$  tests. Association between risk indicators and progressive hearing loss was assessed through logistic regression. The cumulative amount of deterioration in hearing from 1 to 4 years of age was also examined.

**Results:** Our analysis of 330 children (251 exposed to screening) with detailed audiologic records showed that 158 (47.9%) children had some deterioration (at least  $\geq 10$  dB and) in hearing thresholds in at least one ear. The 158 children included 76 (48.1%) with  $\geq 20$  dB loss in pure-tone average in at least one ear and 82 (51.9%) with less deterioration in hearing levels ( $\geq 10$  but  $< 20$  dB). In the children with progressive hearing loss, of 131 children initially diagnosed with bilateral loss, 75 (57.3%) experienced deterioration in 1 ear and 56 (112 ears; 42.7%) in both ears (total of 187 ears). Of 27 children with an initial diagnosis of unilateral loss, 25 experienced deterioration in the impaired ear and 5 in the normal-hearing ear, progressing to bilateral hearing loss. Within 4 years after diagnosis, the mean decrease in hearing for children with progressive loss was 25.9 dB (SD: 16.4) in the right ear and 28.3 dB (SD: 12.9) in the left ear. We explored the risk factors for hearing loss identified by Joint Committee on Infant Hearing where there were sufficient numbers in our sample. On multivariate analysis, there was no statistically significant relationship between most risk indicators examined (neonatal intensive care unit admission, family history, syndromes, and postnatal infections) and the likelihood of progressive loss. However, the presence of craniofacial anomalies was inversely associated with risk of progressive hearing loss (odds ratio = 0.27; 95% confidence interval: 0.10, 0.71;  $p = 0.01$ ), that is, these children were more likely to have stable hearing. **Conclusion:** The results suggest that

the bilirubin induced auditory neuronal damage can be reversible over time so we suggest that infants with hyperbilirubinemia who fail the first hearing tests should be reevaluated after 3 months of treatment.

**Conclusions:** Given that almost half of the children in this cohort experienced deterioration in hearing, close postneonatal monitoring of hearing following early hearing loss identification is essential to ensure optimal amplification and therapy.

[Bass JK, Hua CH, Huang J, Onar-Thomas A, Ness KK, Jones S, White S, Bhagat SP, Chang KW, Merchant TE.](#)  
[Hearing Loss in Patients Who Received Cranial Radiation Therapy for Childhood Cancer.](#)

*J Clin Oncol.* 2016 Apr 10;34(11):1248-55. doi: 10.1200/JCO.2015.63.6738. Epub 2016 Jan 25.

**Purpose:** Patients treated with cranial radiation therapy (RT) are at risk for sensorineural hearing loss (SNHL). Although SNHL is often characterized as a delayed consequence of anticancer therapy, longitudinal reports of SNHL in childhood cancer survivors treated with contemporary RT are limited. We report the incidence, onset, severity, and long-term trajectory of SNHL among children receiving RT. Potential risk factors for SNHL were also identified.

**Patients and Methods:** Serial audiologic testing was conducted on 235 pediatric patients who were treated with conformal or intensity-modulated RT as part of an institutional phase II trial for localized primary brain tumors, including craniopharyngioma, ependymoma, and juvenile pilocytic astrocytoma. All but one patient had measurable cochlear radiation dose (CRD) greater than 0 Gy. The median follow-up from RT initiation to latest audiogram was 9 years with a median of 11 post-RT audiograms per patient. Audiograms were classified by the Chang Ototoxicity Grading Scale. Progression was defined by an increase in Chang grade from SNHL onset to the most recent evaluation.

**Results:** At last evaluation, SNHL was prevalent in 14% of patients: 2.1% had mild and 11.9% had significant SNHL requiring hearing aids. Median time from RT to SNHL onset was 3.6 years (range, 0.4 to 13.2 years). Among 29 patients with follow-up evaluations after SNHL onset, 65.5% experienced continued decline in hearing sensitivity in either ear and 34.5% had no change. Younger age at RT initiation (hazard ratio [HR], 2.32; 95% CI, 1.21 to 4.46), higher CRD (HR, 1.07; 95% CI, 1.03 to 1.11), and cerebrospinal fluid shunting (HR, 2.02; 95% CI, 1.07 to 3.78) were associated with SNHL.

**Conclusions:** SNHL is a late effect of RT that likely worsens over time. Long-term audiologic follow-up for a minimum of 10 years post-RT is recommended.

[Bastanza G, Gallus R, De Carlini M, Picciotti PM, Muzzi E, Ciciriello E, Orzan E, Conti G.](#)  
[Achieving Effective Hearing Aid Fitting within One Month after Identification of Childhood Permanent Hearing Impairment.](#)

*Acta Otorhinolaryngol Ital.* 2016 Feb;36(1):38-44. doi: 10.14639/0392-100X-1077. Epub 2016 Feb 29.

**Abstract:** Diagnosis of child permanent hearing impairment (PHI) can be made with extreme timeliness compared to the past thanks to improvements in PHI identification through newborn hearing screening programmes. It now becomes essential to provide an effective amplification as quickly as possible in order to restore auditory function and favour speech and language development. The early fitting of hearing aids and possible later cochlear implantation indeed prompts the development of central auditory pathways, connections with secondary sensory brain areas, as well as with motor and articulatory cortex. The aim of this paper is to report the results of a strategic analysis that involves identification of strengths, weaknesses, opportunities and threats regarding the process of achieving early amplification in all cases of significant childhood PHI. The analysis is focused on the Italian situation and is part of the Italian Ministry of Health project CCM 2013 "Preventing Communication Disorders: a Regional Program for Early Identification, Intervention and Care of Hearing Impaired Children".

[Bosteels S, Vandenbroeck M, Van Hove G.](#)  
[Saving Deaf Children? Screening for Hearing Loss as a Public-interest Case.](#)

*JJ Bioeth Inq.* 2016 Oct 19.

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

DOI: 10.1007/s11673-016-9752-y

[Boudewyns A, Declau F, van den Ende J, Hofkens A, Dirckx S, Van de Heyning P.](#)  
[Auditory Neuropathy Spectrum Disorder \(ANSD\) in Referrals from Neonatal Hearing Screening at a Well-baby Clinic.](#)

*Eur J Pediatr.* 2016 Jul;175(7):993-1000. doi: 10.1007/s00431-016-2735-5. Epub 2016 May 24.

**Abstract:** Auditory neuropathy spectrum disorder (ANSD) is a particular kind of hearing disorder characterised by normal outer hair cell function and abnormal or absent auditory brain stem responses. Little data are available regarding the prevalence of this condition in healthy newborns. We performed a retrospective medical records review of 791 referrals from universal neonatal hearing screening (UNHS) at a well-baby clinic to investigate the prevalence of ANSD. Hearing screening was performed by automated auditory brain stem response (ABR) testing. A diagnosis of ANSD was established when ABR tracings were absent in the presence of otoacoustic emissions and/or a cochlear microphonic. Amongst 201 infants with confirmed congenital hearing loss, 13 infants were diagnosed with ANSD. The condition was unilateral in six and bilateral in seven infants. A risk factor for hearing loss could be identified in three infants. Abnormalities on magnetic resonance imaging were found in six infants; five of them had cochlear nerve deficiency.

**Conclusion:** The prevalence of ANSD was 6.5 % amongst well babies with confirmed congenital hearing loss identified through UNHS. The estimated incidence of ANSD in our population of newborns at the well-baby clinic was 0.09/1000 live births. Magnetic resonance revealed an underlying anatomical abnormality in about half of the patients.

**What is Known:** Auditory neuropathy dyssynchrony spectrum disorder (ANSD) is a particular form of hearing loss, mostly encountered in neonatal intensive care unit (NICU) graduates. • Little data are available on the prevalence and risk factors for ANSD in healthy newborns. What is new: • The estimated prevalence of ANSD in healthy newborns is 0.09/1000 live births. • In about half of the healthy newborns with ANSD, a structural abnormality was detected on magnetic resonance imaging of the posterior fossa/brain.

[Calcutt TL, Dornan D, Beswick R, Tudehope DI.](#)

### [Newborn Hearing Screening in Queensland 2009-2011: Comparison of Hearing Screening and Diagnostic Audiological Assessment between Term and Preterm Infants.](#)

[J Paediatr Child Health. 2016 Aug 13. doi: 10.1111/jpc.13281.](#)

**AIM:** This study compares rates and timing of newborn hearing screening outcomes, audiological assessment and hearing loss diagnosis between infants of different gestational age groups. Early identification and management of sensorineural hearing loss (SNHL), ideally by 3-6 months of age, facilitates speech and language optimisation. Literature stratifying hearing screening and diagnostic audiology assessment by gestational age groups is lacking.

**Methods:** Subjects were infants with recorded gestational ages receiving newborn hearing screening in Queensland between 2009 and 2011. Data were provided through the Queensland Healthy Hearing database. Infants were analysed in <34 weeks, 34-36+6 weeks, 37-38+6 weeks and ≥39 weeks gestational age groups.

**Results:** Infants (175 911) were eligible for analysis, 7.9% being preterm. Per 1000 infants analysed, bilateral SNHL of >40dB occurred in 2.4 for <34, 1.4 for 34-36+6, 0.7 for 37-38+6 and 0.7 for ≥39 weeks gestation. Diagnoses attributable to newborn hearing screening direct referral were 93.1% for bilateral >40dB SNHL and 88.2% for other hearing loss. Relative to term, preterm infants had a higher incidence of direct and targeted surveillance referrals, audiology assessment and hearing loss diagnosis. Preterm infants were screened later after birth.

**Conclusions:** Specific hearing screening and diagnosis characteristics differed between preterm infants <34 and 34-36+6 weeks gestation, and term infants. Consideration of unique gestational age strata characteristics supports care individualisation. Preterm infants represent a diagnostic challenge, with higher rates of bilateral >40dB SNHL than term but correspondingly higher false positive results on screening, justifying vigilant monitoring. Focused research into specific risk factors in preterm infants is warranted.

[Carey JC, Palumbos JC.](#)

### [Advances in the Understanding of the Genetic Causes of Hearing Loss in Children Inform a Rational Approach to Evaluation. Indian J Pediatr. 2016 Oct;83\(10\):1150-6. doi: 10.1007/s12098-015-1941-x. Epub 2016 Jan 8.](#)

**Abstract:** Hearing loss represents the most common sensory disability of children. Remarkable advances in the identification of genes underlying nonsyndromic and syndromic hearing loss in just the last 2 decades have led to the ability to determine the specific genetic cause of hearing loss in many children. Surprisingly one gene, GJB2, encoding the protein connexin-26, accounts for about 20 % of sensorineural hearing loss (including in India) and is considered the first tier test in evaluating an infant with unexplained congenital hearing loss. Using the knowledge of the etiology of hearing loss, the authors propose a diagnostic reasoning process for the assessment of a child in the pediatric setting. Second tier testing consists of the multiple gene panels using whole exome sequencing strategies, and is becoming available in some regions of the world including the US. Referral to medical genetics is always a consideration in a child with no explanation for the hearing loss and in families with questions about recurrence risk.

[Chiou ST, Lung HL, Chen LS, Yen AM, Fann JC, Chiu SY, Chen HH.](#)

### [Economic Evaluation of Long-term Impacts of Universal Newborn Hearing Screening. Int J Audiol. 2016 Sep 6:1-7.](#)

**Objective:** Little is known about the long-term efficacious and economic impacts of universal newborn hearing screening (UNHS).

**Design:** An analytical Markov decision model was framed with two screening strategies: UNHS with transient evoked otoacoustic emission (TEOAE) test and automatic acoustic brainstem response (aABR) test against no screening. By estimating intervention and long-term costs on treatment and productivity losses and the utility of life years determined by the status of hearing loss, we computed base-case estimates of the incremental cost-utility ratios (ICURs). The scattered plot of ICUR and acceptability curve was used to assess the economic results of aABR versus TEOAE or both versus no screening.

**Study Sample:** A hypothetical cohort of 200,000 Taiwanese newborns.

**Results:** TEOAE and aABR dominated over no screening strategy (ICUR=\$-4800.89 and \$-4111.23, indicating less cost and more utility). Given \$20,000 of willingness to pay (WTP), the probability of being cost-effective of aABR against TEOAE was up to 90%.

**Conclusions:** UNHS for hearing loss with aABR is the most economic option and supported by economically evidence-based evaluation from societal perspective.

**DOI:** 10.1080/14992027.2016.1219777

[Costich JF, Durst AL.](#)

### [The Impact of the Affordable Care Act on Funding for Newborn Screening Services.](#)

[Public Health Rep. 2016 Jan-Feb;131\(1\):160-6.](#)

**Objective:** The Affordable Care Act requires most health plans to cover the federal Recommended Uniform Screening Panel of newborn screening (NBS) tests with no cost sharing. However, state NBS programs vary widely in both the number of mandated tests and their funding mechanisms, including a combination of state laboratory fees, third-party billing, and other federal and state funding. We assessed the potential impact of the Affordable Care Act coverage mandate on states' NBS funding.

**Method:** An analytical Markov decision model was framed with two screening strategies: UNHS with transient evoked otoacoustic emission (TEOAE) test and We performed an extensive review of the refereed literature, federal and state agency reports, relevant organizations' websites, and applicable state laws and regulations; interviewed 28 state and federal officials from August to December 2014; and then assessed the interview findings manually.

**Results:** Although a majority of states had well-established systems for including laboratory-based NBS tests in bundled charges for newborn care, billing practices for critical congenital heart disease and newborn hearing tests were less uniform. Most commonly, birthing facilities either prepaid the costs of laboratory-based tests when acquiring the filter paper kits, or the facilities paid for the tests when the kits were submitted. Some states had separate arrangements for billing Medicaid, and smaller facilities sometimes contracted with hearing test vendors that billed families separately.

**Conclusions:** Although the Affordable Care Act coverage mandate may offset some state NBS funding for the screenings themselves, federal support is still required to assure access to the full range of NBS program services. Limiting reimbursement to the costs of screening tests alone would undermine the common practice of using screening charges to fund follow-up services counseling, and medical food or formula, particularly for low-income families.

[Cubillana-Herrero JD](#), [Pelegrín-Hernández JP](#), [Soler-Valcarcel A](#), [Mínguez-Merlos N](#), [Cubillana-Martínez MJ](#), [Navarro Barrios Á](#), [Medina-Banegas A](#), [Fernandez Hernandez JA](#).

### [The Assessment of the Newborn Hearing Screening Program in the Region of Murcia from 2004 to 2012.](#)

*Int J Pediatr Otorhinolaryngol.* 2016 Sep;88:228-32. doi: 10.1016/j.ijporl.2016.07.009. Epub 2016 Jul 12.

**Objective:** Newborn (NB) auditory deficit has a prevalence of 1-2% in the world. Since the 1990s different screening programs have been put into practice. The Newborn Hearing Screening Program has been in operation since 2002 in our hospital (HCUVA) in Murcia (Spain) and two years later it was introduced into the whole of the Autonomous Community of the Region of Murcia as part of universal healthcare. The objective of this study was to analyze and assess its results.

**Method:** The newborn (NB) population is divided into two groups: not-at-risk NBs and at-risk NBs. In the first case we carry out acoustic otoemissions (AOEs) 48 h after birth and depending on the result the child is either discharged or, in negative cases, the infant undergoes a series of tests in a period of 30-45 days to confirm or rule out the existence of hearing anomalies. In the at-risk group we combine AOEs with brain trunk potentials (BERA) following the stages in a decision tree diagram similar to the ones for not-at-risk children in order to provide a clinical diagnosis in the first three months of life.

**Results:** The screening performance was assessed for the 156,122 children studied, of which 151,258 belonged to the group of not-at-risk children; and 4864 to the group at risk of hypoacusia. As a result of the screening only 410 (0.26%) were sent to consultation, 213 in the not-at-risk group (0.14%) and 197 (24.7%) in the at-risk group. A total of 7452 false positives were identified (4.7%), 6951 (4.5%) in the not-at-risk group; and 501 (10.3%) in the group with risk factors; and there were 53 false negatives (0.03%). Sensitivity in the screening program was 88.5%, with a specificity of 95%.

**Conclusions:** The Region of Murcia has a Newborn Hearing Screening Program with tests that provide a high level of sensitivity and specificity in accordance with the findings of the literature. Our results endorse the program and the patients were treated in a way that met the objective of providing a correct diagnosis and the appropriate therapeutic action.

[de Kock T](#), [Swanepoel D](#), [Hall JW 3rd](#).

### [Newborn Hearing Screening at a Community-based Obstetric Unit: Screening and Diagnostic Outcomes.](#)

*Int J Pediatr Otorhinolaryngol.* 2016 May;84:124-31. doi: 10.1016/j.ijporl.2016.02.031. Epub 2016 Mar 5.

**Objective:** Postnatal visits at community-based midwife obstetric units (MOUs) have been proposed as an alternative primary healthcare screening platform in South Africa. This study evaluated the outcomes of distortion product otoacoustic emissions (DPOAEs) and automated auditory brainstem response (AABR) screening conducted by a dedicated non-professional screener at a community-based MOU in the Western Cape, South Africa.

**Method:** Universal newborn hearing screening (UNHS) at a community-based MOU was evaluated over a 16-month period. A dedicated non-professional screener was trained to follow a two-stage screening protocol targeting bilateral hearing loss. A two group comparative design was used alternating AABR (Maico MB11 BERAPHONE™()) and DPOAE (Bio-logic AuDX I) technology on a daily basis. Infants referring the initial screen received a follow-up appointment in two days' time and were rescreened with the same technology used at their first screen. Those referring the second stage were booked for diagnostic assessments.

**Results:** 7452 infants were screened including 47.9% (n=3573) with DPOAE and 52.1% (n=3879) with AABR technology. Mean age at first stage screen was 6.1 days. The initial bilateral referral rate was significantly lower for AABR (4.6%) compared to DPOAE (7.0%) and dropped to 0.3% and 0.7% respectively following the second stage screenings. First rescreen and initial diagnostic follow-up rates of 90% and 92.3% were obtained for the DPOAE group and 86.6% and 90% for the AABR group. Follow-up rates showed no significant difference between technology groups. Diagnostic assessment revealed a higher prevalence rate for bilateral SNHL among the AABR group (1/1000) compared to the DPOAE group (0.3/1000). Screening technology had no significant influence on daily screening capacity (23 AABR/day; 24 DPOAE/day).

**Conclusions:** Postnatal visits at community-based MOUs create a useful platform for hearing screening and follow-up. AABR technology with negligible disposable costs provides opportunity for AABR screening to be utilised in community-based programmes. AABR screening offers lower initial referral rates and a higher true positive rate compared to DPOAE.

[Dettman S](#), [Choo D](#), [Dowell R](#).

### [Barriers to Early Cochlear Implantation.](#)

*Int J Audiol* 2016;55 Suppl 2:S64-76. doi: 10.1080/14992027.2016.1174890. Epub 2016 May 3.

**Objective:** Identify variables associated with paediatric access to cochlear implants (CIs).

**Design:** Part 1. Trends over time for age at CI surgery (N=802) and age at hearing aid (HA) fitting (n=487) were examined with regard to periods before, during, and after newborn hearing screening (NHS). Part 2. Demographic factors were explored for 417 children implanted under 3 years of age. Part 3. Pre-implant steps for the first 20 children to receive CIs under 12 months were examined.

**Results:** Part 1. Age at HA fitting and CI surgery reduced over time, and were associated with NHS implementation. Part 2. For children implanted under 3 years, earlier age at HA fitting and higher family socio-economic status were associated with earlier CI. Progressive hearing loss was associated with later CIs. Children with a Connexin 26 diagnosis received CIs earlier than children with a premature / low birth weight history. Part 3. The longest pre-CI steps were Step 1: Birth to diagnosis/identification of hearing loss (mean 16.43 weeks), and Step 11: MRI scans to implant surgery (mean 15.05 weeks) for the first 20 infants with CIs under 12 months.

**Conclusions:** NHS implementation was associated with reductions in age at device intervention in this cohort.

[Elpers J](#), [Lester C](#), [Shinn JB](#), [Bush ML](#).

### [Rural Family Perspectives and Experiences with Early Infant Hearing Detection and Intervention: A Qualitative Study.](#) *J Community Health.* 2016 Apr;41(2):226-33. doi: 10.1007/s10900-015-0086-1.

**Abstract:** Infant hearing loss has the potential to cause significant communication impairment. Timely diagnosis and intervention is essential to preventing permanent deficits. Many infants from rural regions are delayed in diagnosis and treatment of hearing loss. The purpose of this study is to characterize the barriers in timely infant hearing healthcare for rural families following newborn newborn hearing screening (NHS) testing. Using stratified purposeful sampling, the study design involved semi-structured phone interviews with parents/guardians of children who failed NHS testing in the Appalachian region of Kentucky between 2012 and 2014 to describe their experiences with early hearing detection and intervention program. Thematic qualitative analysis was performed on interview transcripts to identify common recurring themes in content. 40 parents/guardians participated in the study and consisted primarily of mothers. Demographic data revealed limited educational levels of the participants and 70 % had state-funded insurance coverage. Participants reported barriers in timely infant hearing healthcare that included poor communication of hearing screening results, difficulty in obtaining outpatient testing, inconsistencies in healthcare information from primary care providers, lack of local resources, insurance-related healthcare delays, and conflict with family and work responsibilities. Most participants expressed a great desire to obtain timely hearing healthcare for their children and expressed a willingness to use resources such as telemedicine to obtain that care. There are multiple barriers to timely rural infant hearing healthcare. Minimizing misinformation and improving access to care are priorities to prevent delayed diagnosis

[Farzal Z](#), [Kou YF](#), [St John R](#), [Shah GB](#), [Mitchell RB](#).

### [The Role of Routine Hearing Screening in Children with Cystic Fibrosis on Aminoglycosides: A Systematic Review.](#)

*Laryngoscope*. 2016 Jan;126(1):228-35. doi: 10.1002/lary.25409. Epub 2015 Jul 7.

**Objective:** To review the role of routine hearing screening for sensorineural hearing loss (SNHL) in children with cystic fibrosis (CF) who have been on aminoglycoside therapy.

**Data Sources:** PubMed, Cochrane, Scopus, and Ovid databases.

**Review Methods:** A systematic review of the literature was performed in accordance with PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines. A comprehensive search was performed from 1970 to 2014. Randomized controlled trials, case-control studies, cohort studies, and case series including pediatric subjects with baseline auditory evaluations were included.

**Results:** Twelve studies (1979-2014) were reviewed. The study population included 762 children (5 months-20 years). Hearing screening measures included pure-tone audiometry (PTA) at standard  $\pm$  high frequency threshold (HFPTA) (12/12), distortion product otoacoustic emissions (DPOAE) (4/12), transient-evoked otoacoustic emissions (1/12), and automated auditory brainstem response (1/12). The overall prevalence of SNHL ranged from 0% to 29%. However, on subset analysis of children with greater than 10 courses of intravenous (IV) aminoglycosides, up to 44% had SNHL. Eight studies recommended hearing screening in CF children on aminoglycosides; of these, two studies recommended screening even without aminoglycoside exposure, and four studies made no recommendations. HFPTA was the most commonly recommended screening measure followed by DPOAEs.

**Conclusions:** This systematic review supports a recommendation for clinicians to perform routine hearing screening in children with CF during and after aminoglycoside exposure based on the high prevalence of SNHL in this population. Future studies should define the optimal timing for hearing screening during and after aminoglycoside therapy in children with CF.

[Fitzpatrick E](#), [Grandpierre V](#), [Durieux-Smith A](#), [Gaboury I](#), [Coyle D](#), [Na E](#), [Sallam N](#).

### [Children With Mild Bilateral and Unilateral Hearing Loss: Parents' Reflections on Experiences and Outcomes.](#)

*J Deaf Stud Deaf Educ*. 2016 Jan;21(1):34-43. doi: 10.1093/deafed/env047. Epub 2015 Oct 3.

**Abstract:** Children with mild bilateral and unilateral hearing loss are now commonly identified early through newborn hearing screening initiatives. There remains considerable uncertainty about how to support parents and about which services to provide for children with mild bilateral and unilateral hearing loss. The goal of this study was to learn about parents' experiences and understand, from their perspectives, the impact of hearing loss in the mild range on the child's functioning. Parents of 20 children in Ontario, Canada, participated in the study. The median age of identification of hearing loss was 4.6 months (interquartile range: 3.6, 10.8). Parents appreciated learning early about hearing loss, but their experiences with the early process were mixed. Parents felt that professionals minimized the importance of milder hearing loss. There was substantial uncertainty about the need for hearing aids and the findings suggest that parents need specific guidance. Parents expressed concerns about the potential impact of hearing loss on their child's development, particularly at later ages.

[Fitzpatrick EM](#), [Hamel C](#), [Stevens A](#), [Pratt M](#), [Moher D](#), [Doucet SP](#), [Neuss D](#), [Bernstein A](#), [Na E](#).

### [Sign Language and Spoken Language for Children With Hearing Loss: A Systematic Review.](#)

*Pediatrics*. 2016 Jan;137(1). doi: 10.1542/peds.2015-1974. Epub 2015 Dec 18.

**Context:** Permanent hearing loss affects 1 to 3 per 1000 children and interferes with typical communication development. Early detection through newborn hearing screening and hearing technology provide most children with the option of spoken language acquisition. However, no consensus exists on optimal interventions for spoken language development.

**Objective:** To conduct a systematic review of the effectiveness of early sign and oral language intervention compared with oral language intervention only for children with permanent hearing loss.

**Data Sources:** An a priori protocol was developed. Electronic databases (eg, Medline, Embase, CINAHL) from 1995 to June 2013 and gray literature sources were searched. Studies in English and French were included.

**Study Selection:** Two reviewers screened potentially relevant articles.

**Data Extraction:** Outcomes of interest were measures of auditory, vocabulary, language, and speech production skills. All data collection and risk of bias assessments were completed and then verified by a second person. Grades of Recommendation, Assessment, Development, and Evaluation (GRADE) was used to judge the strength of evidence.

**Results:** Eleven cohort studies met inclusion criteria, of which 8 included only children with severe to profound hearing loss with cochlear implants. Language development was the most frequently reported outcome. Other reported outcomes included speech and speech perception.

**Limitations:** Several measures and metrics were reported across studies, and descriptions of interventions were sometimes unclear.

**Conclusions:** Very limited, and hence insufficient, high-quality evidence exists to determine whether sign language in combination with oral language is more effective than oral language therapy alone. More research is needed to supplement the evidence base.

[Fortnum H](#), [Ukoumunne OC](#), [Hyde C](#), [Taylor RS](#), [Ozolins M](#), [Errington S](#), [Zhelev Z](#), [Pritchard C](#), [Benton C](#), [Moody J](#), [Cocking L](#), [Watson J](#), [Roberts S](#).

### [A Programme of Studies including Assessment of Diagnostic Accuracy of School Hearing Screening Tests and a Cost-effectiveness Model of School Entry Hearing Screening Programmes.](#)

*Health Technol Assess*. 2016 May;20(36):1-178. doi: 10.3310/hta20360.

**Background:** Identification of permanent hearing impairment at the earliest possible age is crucial to maximise the development of speech and language. Universal newborn hearing screening identifies the majority of the 1 in 1000 children born with a hearing impairment, but later onset can occur at any time and there is no optimum time for further screening. A universal but non-standardised school entry screening (SES) programme is in place in many parts of the UK but its value is questioned.

**Objectives:** To evaluate the diagnostic accuracy of hearing screening tests and the cost-effectiveness of the SES programme in the UK.

**Design:** Systematic review, case-control diagnostic accuracy study, comparison of routinely collected data for services with and without a SES programme, parental questionnaires, observation of practical implementation and cost-effectiveness modelling.

**Setting:** Second- and third-tier audiology services; community.

**Participants:** Children aged 4-6 years and their parents.

**Main Outcome Measures:** Diagnostic accuracy of two hearing screening devices, referral rate and source, yield, age at referral and cost per quality-adjusted life-year.

**Results:** The review of diagnostic accuracy studies concluded that research to date demonstrates marked variability in the design, methodological quality and results. The pure-tone screen (PTS) (Amplivox, Eynsham, UK) and HearCheck (HC) screener (Siemens, Frimley, UK) devices had high

sensitivity (PTS  $\geq$ 89%, HC  $\geq$ 83%) and specificity (PTS  $\geq$ 78%, HC  $\geq$ 83%) for identifying hearing impairment. The rate of referral for hearing problems was 36% lower with SES (Nottingham) relative to no SES (Cambridge) [rate ratio 0.64, 95% confidence interval (CI) 0.59 to 0.69;  $p < 0.001$ ]. The yield of confirmed cases did not differ between areas with and without SES (rate ratio 0.82, 95% CI 0.63 to 1.06;  $p = 0.12$ ). The mean age of referral did not differ between areas with and without SES for all referrals but children with confirmed hearing impairment were older at referral in the site with SES (mean age difference 0.47 years, 95% CI 0.24 to 0.70 years;  $p < 0.001$ ). Parental responses revealed that the consequences to the family of the referral process are minor. A SES programme is unlikely to be cost-effective and, using base-case assumptions, is dominated by a no screening strategy. A SES programme could be cost-effective if there are fewer referrals associated with SES programmes or if referrals occur more quickly with SES programmes.

**Conclusions:** A SES programme using the PTS or HC screener is unlikely to be effective in increasing the identified number of cases with hearing impairment and lowering the average age at identification and is therefore unlikely to represent good value for money. This finding is, however, critically dependent on the results of the observational study comparing Nottingham and Cambridge, which has limitations. The following are suggested: systematic reviews of the accuracy of devices used to measure hearing at school entry; characterisation and measurement of the cost-effectiveness of different approaches to the ad-hoc referral system; examination of programme specificity as opposed to test specificity; further observational comparative studies of different programmes; and opportunistic trials of withdrawal of SES programmes.

[Gantt S, Dionne F, Kozak FK, Goshen O, Goldfarb DM, Park AH, Boppana SB, Fowler K.](#)  
[Cost-effectiveness of Universal and Targeted Newborn Screening for Congenital Cytomegalovirus Infection.](#)  
[JAMA Pediatr.](#) 2016 Oct 10. doi: 10.1001/jamapediatrics.2016.2016.

**Importance:** Congenital cytomegalovirus (cCMV) infection is a major cause of childhood deafness. Most cCMV infections are not diagnosed without newborn screening, resulting in missed opportunities for directed care.

**Objective:** To estimate the cost-effectiveness of universal and targeted newborn cCMV screening programs compared with no cCMV screening.

**Design, Setting, and Participants:** Models were constructed using rates and outcomes from prospective cohort studies of newborn cCMV screening in US postpartum care and early hearing programs. Costs of laboratory testing, treatment, and hearing loss were drawn from Medicaid data and published estimates. The benefits of cCMV screening were assumed to come from antiviral therapy for affected newborns to reduce hearing loss and from earlier identification of hearing loss with postnatal onset. Analyses were performed from July 2014 to March 2016.

**Interventions:** Models compared universal or targeted cCMV screening of newborns with a failed hearing screen, with standard care for cCMV infection.

**Main Outcome and Measures:** The incremental costs of identifying 1 cCMV infection, identifying 1 case of cCMV-related hearing loss, and preventing 1 cochlear implant; the incremental reduction in cases of severe to profound hearing loss; and the differences in costs per infant screened by universal or targeted strategies under different assumptions about the effectiveness of antiviral treatment.

**Results:** Among all infants born in the United States, identification of 1 case of cCMV infection by universal screening was estimated to cost \$2000 to \$10 000; by targeted screening, \$566 to \$2832. The cost of identifying 1 case of hearing loss due to cCMV was as little as \$27 460 by universal screening or \$975 by targeted screening. Assuming a modest benefit of antiviral treatment, screening programs were estimated to reduce severe to profound hearing loss by 4.2% to 13% and result in direct costs of \$10.86 per newborn screened. However, savings of up to \$37.97 per newborn screened were estimated when costs related to functionality were included.

**Conclusions and Relevance:** Newborn screening for cCMV infection appears to be cost-effective under a wide range of assumptions. Universal screening offers larger net savings and the greatest opportunity to provide directed care. Targeted screening also appears to be cost-effective and requires testing for fewer newborns. These findings suggest that implementation of newborn cCMV screening programs is warranted.

[Gardner-Berry K, Chang H, Ching TY, Hou S.](#)  
[Detection Rates of Cortical Auditory Evoked Potentials at Different Sensation Levels in Infants with Sensory/Neural Hearing Loss and Auditory Neuropathy Spectrum Disorder.](#)  
[Semin Hear.](#) 2016 Feb;37(1):53-61. doi: 10.1055/s-0035-1570330.

**Abstract:** With the introduction of newborn hearing screening, infants are being diagnosed with hearing loss during the first few months of life. For infants with a sensory/neural hearing loss (SNHL), the audiogram can be estimated objectively using auditory brainstem response (ABR) testing and hearing aids prescribed accordingly. However, for infants with auditory neuropathy spectrum disorder (ANSD) due to the abnormal/absent ABR waveforms, alternative measures of auditory function are needed to assess the need for amplification and evaluate whether aided benefit has been achieved. Cortical auditory evoked potentials (CAEPs) are used to assess aided benefit in infants with hearing loss; however, there is insufficient information regarding the relationship between stimulus audibility and CAEP detection rates. It is also not clear whether CAEP detection rates differ between infants with SNHL and infants with ANSD. This study involved retrospective collection of CAEP, hearing threshold, and hearing aid gain data to investigate the relationship between stimulus audibility and CAEP detection rates. The results demonstrate that increases in stimulus audibility result in an increase in detection rate. For the same range of sensation levels, there was no difference in the detection rates between infants with SNHL and ANSD.

[Giuntini G, Forli F, Nicastro R, Ciabotti A, Bruschini L, Berrettini S.](#)  
[Early Care in Children with Permanent Hearing Impairment.](#)  
[Acta Otorhinolaryngol Ital.](#) 2016 Feb;36(1):51-9. doi: 10.14639/0392-100X-1079. Epub 2016 Feb 29.

**Abstract:** The implementation of regional protocols for newborn hearing screening and early audiologic diagnosis represent the first step of the entire diagnostic, rehabilitative and prosthetic programme for children with permanent hearing impairment. The maximum benefit of early diagnosis can indeed be obtained only by prompt rehabilitation aimed at fostering the child's communicative, linguistic and cognitive development. Within the framework of the CMM 2013 project of the Ministry of Health entitled "Preventing Communication Disorders: a Regional Program for Early Identification, Intervention and Care of Hearing Impaired Children", the problems concerning the promotion of the global development of children with PHI through an early rehabilitation project based on shared knowledge and scientific evidence. In this project, our specific aim was to define the features and modes of access to a precise and specialised rehabilitation project for the small hearing-impaired child within three months from audiologic diagnosis. Three main recommendations relative to assessment and rehabilitation aspects of early care emerged from the study.

[Holzinger D, Weishaupt A, Fellingner P, Beitel C, Fellingner J.](#)  
[Prevalence of 2.2 Per Mille of Significant Hearing Loss at School Age Suggests Rescreening After NHS.](#)  
[Int J Pediatr Otorhinolaryngol.](#) 2016 Aug;87:121-5. doi: 10.1016/j.ijporl.2016.06.006. Epub 2016 Jun 7.

**Objectives:** The study aims to determine the prevalence of different degrees of significant hearing loss in a complete sample of Austrian school-age children born between 1997 and 2001 living in the federal state of Carinthia and to evaluate the role of Newborn Hearing Screening (NHS) in the identification of later hearing loss.

**Methods:** In Carinthia, all school-age children with significant hearing loss (mean pure tone average in the better ear above 40 dB) are registered by the Department of Education. From five complete birth cohorts from 1997 to 2001 (n = 28.171) all the children with sensorineural hearing loss (n = 61, mean age 10.5, age range 7.5-13.6 years) were assessed for their hearing threshold and level of cognitive functioning. Socio-demographic data, including information about NHS and amplification with hearing devices, were collected from parents and teachers using structured interviews.

**Results:** 2.2 children per thousand (49.2% male) were found to be affected by significant bilateral hearing loss at school age, with 36.1% of them having a moderate hearing loss, 34.4% severe, and 29.5% profound. Fourteen children (23.0%) used cochlear implants. Their mean nonverbal IQ was 93.4 (SD 23.1), including 13.1% of children with intellectual disabilities (IQ < 70). Of those who had undergone NHS (85.2%), 50.0% had passed the screening according to parents' reports.

**Conclusion:** A rate of significant hearing loss in school-age children was observed which was twice the rate found in newborns. Ongoing awareness of late-onset hearing loss to improve identification and hearing screening at school entry are recommended.

[Hu T, Stead K, Fu T, Papsin B.](#)

### [A Program Evaluation of Kids2Hear, a Student-run Hearing Screening Program for School Children.](#)

[J Otolaryngol Head Neck Surg.](#) 2016 Sep 26;45(1):49.

**Background:** Hearing deficits in children are demonstrably negatively associated with language acquisition and cognition. Although universal neonatal hearing screening exists, it is not offered equally across Canada. Additionally, children emigrating from other countries are often not assessed. The objective of this study is to evaluate Kids2Hear, a free hearing screening program run by medical students at elementary schools, and to determine the rate of hearing deficits that were identified and referred for evaluation.

**Methods:** Retrospective analysis of screening program data from 228 participants seen at three inner-city elementary schools over six months.

**Results:** In our sample, the mean age was 5.8 ± 1.0 years with 48 % males. Approximately 21 participants (9.3 %) were screened positive for a hearing deficit and required referral for supplementary audiological evaluation. About 44 participants (19.3 %) were referred to a family physician for otoscopic abnormalities. Females were significantly more likely to be identified for both hearing deficits and otoscopic abnormalities.

**Conclusions:** Hearing deficits and otoscopic abnormalities are common among young children. Female children may be at higher risk for developing hearing issues or otoscopic abnormalities compared to males. Additional research is needed to determine the effectiveness of hearing screening programs.

DOI: 10.1186/s40463-016-0159-x

[Hunter LL, Keefe DH, Feeney MP, Fitzpatrick DF, Lin L.](#)

### [Longitudinal Development of Wideband Reflectance Tympanometry in Normal and At-risk Infants.](#)

[Hear Res.](#) 2016 Oct;340:3-14. doi: 10.1016/j.heares.2015.12.014. Epub 2015 Dec 19.

**Purpose:** The goals of this study were to measure normal characteristics of ambient and tympanometric wideband acoustic reflectance, which was parameterized by absorbance and group delay, in newborns cared for in well-baby and Neonatal Intensive Care Unit (NICU) nurseries, and to characterize the normal development of reflectance over the first year after birth in a group of infants with clinically normal hearing status followed longitudinally from birth to one year of age.

**Methods:** Infants were recruited from a well-baby and NICU nursery, passed newborn otoacoustic emissions (OAE) and automated auditory brainstem response (ABR) tests as well as follow-up diagnostic ABR and audiometry. They were tested longitudinally for up to one year using a wideband middle ear acoustic test battery consisting of tympanometry and ambient-pressure tests. Results were analyzed for ambient reflectance across frequency and tympanometric reflectance across frequency and pressure.

**Results:** Wideband absorbance and group delay showed large effects of age in the first 6 months. Immature absorbance and group delay patterns were apparent in the low frequencies at birth and one month, but changed substantially to a more adult-like pattern by age 6 months for both ambient and tympanometric variables. Area and length of the ear canal estimated acoustically increased up to age 1 year. Effects of race (African American and others compared to Caucasian) were found in combination with age effects. Mean and confidence intervals are provided for use as a normative longitudinal database for newborns and infants up to one year of age, for both well-baby and NICU infants.

[Hunter LL, Meinzen-Derr J, Wiley S, Horvath CL, Kothari R, Wexelblatt S.](#)

### [Influence of the WIC Program on Loss to Follow-up for Newborn Hearing Screening.](#)

[Pediatrics.](#) 2016 Jul;138(1). pii: e20154301. doi: 10.1542/peds.2015-4301. Epub 2016 Jun 15.

**Background:** Newborn hearing screening has a high participation rate of ~97% of infants nationally, but a high lost to follow-up of ~32% limits the effectiveness of the program. This study tested an intervention of targeted outpatient rescreening of infants through collaboration with the Women, Infants, and Children (WIC) program to improve follow-up rates for newborn hearing screen referrals.

**Methods:** Controlled intervention study of WIC-eligible infants who referred on newborn hearing screens at target hospitals. Hearing rescreens were performed by using screening auditory brainstem response testing by trained research assistants, coordinated with the infant's WIC appointment. Loss to follow-up rates and age at follow-up were compared with non-WIC infants tracked via the Ohio Department of Health during the same time periods at the same hospitals and at nonintervention hospitals.

**Results:** During a 2-year period, there were 1493 hearing screen referrals at 6 hospitals in the Cincinnati region recorded by the Ohio Department of Health. Of these, 260 WIC-eligible infants were referred to the study. Among WIC-eligible intervention infants, the lost to follow-up rate over 2 years was 9.6%, compared with 28.7% for nonintervention infants in the same hospitals and 18.1% for nonintervention hospitals. The average age of hearing confirmation for the WIC intervention group was 34.8 days, compared with 63.6 days in non-WIC infants. One-third of mothers reported barriers to follow-up.

**Conclusions:** Collaborating with WIC to provide targeted follow-up for newborn hearing screening improved loss to follow-up rates, decreased the age at hearing confirmation by 1 month, and addressed reported care barriers.

[Jeong SW, Kang MY, Kim JR, Kim LS.](#)

### [Delayed-onset Hearing Loss in Pediatric Candidates for Cochlear Implantation.](#)

[Eur Arch Otorhinolaryngol.](#) 2016 Apr;273(4):879-87. doi: 10.1007/s00405-015-3646-1. Epub 2015 May 9.

**Abstract:** The objective of this study was to evaluate the clinical significance of delayed-onset hearing loss in children. Seventy-three children who underwent cochlear implantation (CI) were included. They were divided into a congenital hearing loss group (n = 50) and a delayed-onset hearing loss group (n = 23). The age at diagnosis of hearing loss, age at the beginning of auditory habilitation, the age at CI, and the postimplant speech perception abilities were compared between the two groups. Children in the congenital hearing loss group were confirmed to have hearing loss at a mean age of 0.3 years, and those in the delayed-onset hearing loss group were diagnosed with hearing loss at a mean age of 2.0 years. Auditory habilitation began at a mean age of 0.4 and 2.0 years, and CI was performed at a mean age of 1.4 and 2.6 years, respectively. Children in the congenital hearing loss group had better scores on speech perception tests than those in the delayed-onset hearing loss group, but the differences

were not significant. About half of the children with delayed-onset hearing loss (57 %) had risk factors associated with delayed-onset hearing loss. A high prevalence of delayed-onset hearing loss was noted in the group of children who underwent CI. Risk factors for hearing loss were not found in 43 % of children with delayed-onset hearing loss. Universal screening for delayed-onset hearing loss needs to be performed during early childhood.

[Jiang ZD, Xu ZM, Wilkinson AR.](#)

### [Comparison of Maturation Process of Hearing Threshold in Early Life Between At-risk and Low-risk Preterm Infants.](#)

[Early Hum Dev. 2016 May;96:21-5. doi: 10.1016/j.earlhumdev.2016.02.007. Epub 2016 Mar 15.](#)

**AIM:** To detect any abnormality in the maturational process of hearing threshold during the early life in at-risk preterm infants.

**Study Design:** The threshold of brainstem auditory evoked response was recorded and analyzed longitudinally from 30 to 42 weeks of postconceptional age in 357 at-risk infants born at 23-36 weeks of gestation. The results were compared with those in 82 low-risk infants born at 30-42 weeks at various postconceptional ages.

**Results:** From 31 to 42 weeks, the response threshold in the at-risk infants was consistently slightly higher than that in the low-risk infants. No statistically significant difference was found between the two groups of infants at any designated postconceptional ages. The threshold in the at-risk infants born at 23-29 weeks of gestation tended to be higher than those born at 30-36 weeks at various postconceptional ages, but the difference did not reach statistical significance. There was also no significant difference in the slope of BAER threshold-age function between the at-risk infants, irrespective of gestational ages, and the low-risk infants.

**Conclusions:** During the early life, hearing threshold in at-risk preterm, mainly very preterm, infants is marginally elevated, but the maturational process of the threshold is generally similar to that in low-risk infants, without notable abnormality.

[Kaga K.](#)

### [Auditory Nerve Disease and Auditory Neuropathy Spectrum Disorders.](#)

[Auris Nasus Larynx. 2016 Feb;43\(1\):10-20. doi: 10.1016/j.anl.2015.06.008. Epub 2015 Jul 22.](#)

**Abstract:** In 1996, a new type of bilateral hearing disorder was discerned and published almost simultaneously by Kaga et al. [1] and Starr et al. [2]. Although the pathophysiology of this disorder as reported by each author was essentially identical, Kaga used the term "auditory nerve disease" and Starr used the term "auditory neuropathy". Auditory neuropathy (AN) in adults is an acquired disorder characterized by mild-to-moderate pure-tone hearing loss, poor speech discrimination, and absence of the auditory brainstem response (ABR) all in the presence of normal cochlear outer hair cell function as indicated by normal distortion product otoacoustic emissions (DPOAEs) and evoked summing potentials (SPs) by electrocochleography (ECoG). A variety of processes and etiologies are thought to be involved in its pathophysiology including mutations of the OTOF and/or OPA1 genes. Most of the subsequent reports in the literature discuss the various auditory profiles of patients with AN [3,4] and in this report we present the profiles of an additional 17 cases of adult AN. Cochlear implants are useful for the reacquisition of hearing in adult AN although hearing aids are ineffective. In 2008, the new term of Auditory Neuropathy Spectrum Disorders (ANSO) was proposed by the Colorado Children's Hospital group following a comprehensive study of newborn hearing test results. When ABRs were absent and DPOAEs were present in particular cases during newborn screening they were classified as ANSO. In 2013, our group in the Tokyo Medical Center classified ANSO into three types by following changes in ABRs and DPOAEs over time with development. In Type I, there is normalization of hearing over time, Type II shows a change into profound hearing loss and Type III is true auditory neuropathy (AN). We emphasize that, in adults, ANSO is not the same as AN.

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

### [Feasibility of Newborn Hearing Screening in a Public Hospital Setting in South Africa: A Pilot Study.](#)

[S Afr J Commun Disord. 2016;63\(1\). doi: 10.4102/sajcd.v63i1.150.](#)

**Objectives:** The current pilot study aimed to explore the feasibility of newborn hearing screening (NHS) in a hospital setting with clinical significance for the implementation of NHS. Context-specific Objectives included determining the average time required to screen each neonate or infant; the most suitable time for initial hearing screening in the wards; as well as the ambient noise levels in the wards and at the neonatal follow-up clinic where screening would be conducted.

**Method:** A descriptive, longitudinal, repeated measures, within-subjects design was employed. The pilot study comprised 11 participants who underwent hearing screening. Data were analysed using descriptive statistics.

**Results:** The average time taken to conduct hearing screening using otoacoustic emissions and automated auditory brainstem response was 18.4 minutes, with transient evoked otoacoustic emissions taking the least time. Ambient noise levels differed between wards and clinics with the sound level readings ranging between 50 dBA and 70 dBA. The most suitable screening time was found to be the afternoons, after feeding times.

**Conclusions:** Findings highlight important considerations when embarking on larger scale NHS studies or when planning a hospital NHS programme. Current findings suggest that NHS can be efficiently and effectively conducted in public sector hospitals in South Africa, provided that test time is considered in addition to sensitivity and specificity when deciding on a screening protocol; bar recognised personnel challenges.

[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 Neonatol. 2016 Aug 9. pii: S1875-9572\(16\)30102-4. doi: 10.1016/j.pedneo.2016.05.003.](#)

**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  $\leq 32$  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.

**Conclusions:** 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.



[Kocoń S, Wiatr M, Stręk P, Wiatr A, Grudzień-Ziarno A, Hartwich P.](#)

### [Analysis of Difficulties Occurring During the Early Auditory Screening in Children.](#)

*Otolaryngol Pol.* 2016 Jun 30;70(4):41-8. doi: 10.5604/00306657.1202780.

**Introduction:** It is assumed that the critical period for diagnosis of hearing disorders is the baby's first three months of life and that appropriate course and implementation of treatment and/or rehabilitation should begin before a child is six months old. However various kinds of problems may occur during auditory screening of a child may exceed this interval. This problem is particularly pronounced among children with development and health problems and leads to unreliable and varied results.

**AIM:** The aim of this study was an analysis of prevalence of difficulties occurring during the first year of auditory screening among groups of children with congenital hearing impairment.

**Material and Methods:** Patients were examined in The Universal Newborn Hearing Screening Program in the years 2012 - 2013 in Level III NICUs in Krakow. Results from 250 cases were analyzed retrospectively. Medical exam results of patients with high risk of hearing loss were also included in our analysis. The groups of children included in our study were: children with Down Syndrome, children with nervous system disorders, children with cleft palate or both cleft palate and lip and children with congenital CMV.

**Results:** In the group of children with cleft palate or both cleft palate and lip the most frequent cause of not conducting objective audiometric tests was bad health condition of a child which precluded his arrival for administering the tests. The most common cause of difficulties in performing hearing tests was the emotional state of children from groups with Down Syndrome. In the group of children with congenital CMV the most common cause of difficulties was a lack of availability of their parents.

**Conclusions:** 1. We encountered the greatest diagnostic difficulties during the child's first year of life in chosen high-risk groups of children with congenital hearing loss in children with cleft palate or both cleft palate and lip. 2. The highest prevalence of not finished tests was in III and IV interval for all chosen high-risk groups with congenital hearing loss.

[Li PC, Chen WI, Huang CM, Liu CJ, Chang HW, Lin HC.](#)

### [Comparison of Newborn Hearing Screening in Well-Baby Nursery and NICU: A Study Applied to Reduce Referral Rate in NICU.](#)

*PLoS One.* 2016 Mar 29;11(3):e0152028. doi: 10.1371/journal.pone.0152028. eCollection 2016.

**Objectives:** To determine whether newborn hearing screening in a well-baby nursery (WBN) and neonatal intensive care unit (NICU) nursery: 1) meet three targeted, screening, referral, and diagnostic follow-up rates; 2) compare the average age of diagnosis for infants admitted to the WBN and NICU; and 3) determine prevalence of hearing loss in neonatal population; and 4) try to find a practical newborn hearing screening time algorithm to reduce refer rate in NICU.

**Material and Methods:** The screening rates were 99.8% and 99.6% in the WBN and NICU groups, respectively, without significant difference. The referral rates were 0.7% and 2.8% in the WBN and NICU groups, with significant difference. Furthermore, the diagnostic follow-up rates were 76.7% and 89.1% in the WBN and NICU groups, without significant difference. The average initial diagnostic ages were 1.9 months and 3.8 months in the WBN and NICU groups, with significant difference. The prevalence of congenital bilateral hearing loss were 0.27% and 1.6% in the WBN and NICU groups, with significant difference.

**Conclusions:** The screening, referral and follow-up rate in the WBN and NICU groups were equivalent to the quality indicators. For NICU group, screening and diagnostic follow up were performed later than those in WBN group; however the lower referral rate in our NICU group was successfully achieved in this study and can be applied clinically. The prevalence of congenital bilateral hearing loss was higher in the NICU group than in the WBN group.

[Luz I, Ribas A, Kozłowski L, Willig M, Berberian AP.](#)

### [Newborn Hearing Screening in a Public Maternity Ward in Curitiba, Brazil: Determining Factors for Not Retesting.](#)

*Int Arch Otorhinolaryngol.* 2016 Oct;20(4):300-304. Epub 2015 Nov 16.

**Introduction:** Law 12.303/10 requires hearing screening in newborns before hospital discharge to detect possible hearing problems within the first three months after birth. If the newborn fails the test or presents signs of risk for hearing loss, it must undergo a retest and monitoring during the first year of life. In practice, this often does not happen.

**Objective:** To identify, in a group of mothers of children with risk factors for hearing loss, the determining reasons for non-compliance with the auditory retest.

**Method:** This is a cross-sectional quantitative study. For data collection, we handed a semi-structured questionnaire to 60 mothers of babies at risk for hearing loss who did not attend the hearing retest after hospital discharge. The questionnaire investigated their age, education, marital status, level of knowledge about the hearing screening, and reasons for non-compliance with the retest. We compared and analyzed data using the Chi-square test at a significance level of 0.05%.

**Results:** Our study found that 63% of the respondents were unaware of the hearing screening and most did not receive guidance on testing during prenatal care; 30% of participants stated forgetting as the reason for not attending the retest. There was no significant relationship between age, education, and marital status regarding knowledge about the test and the non-compliance with the retest.

**Conclusion:** Identified as the most significant determining factors for non-compliance with the newborn hearing screening retest were the surveyed mothers' forgetting the date, and their ignorance as to the importance of retesting.

[Martínez-Pacheco MC, Ferrán de la Cierva L, García-Purriños FJ.](#)

### [Delayed Diagnosis of Childhood Deafness: The Value of False Negatives in the Programme for Early Detection of Neonatal Hearing Loss.](#)

*Acta Otorrinolaringol Esp.* 2016 Apr 6. pii: S0001-6519(16)00042-X. doi: 10.1016/j.otorri.2016.01.004.

**Introduction:** Despite its importance, the existence of false negatives (patients who are told they hear well, but they have some degree of hipacusia) is rarely evaluated in programs for early detection of hearing loss. The aim of this study is to determine the variables that can lead to a delayed diagnosis, especially the existence of false negatives and the lack of registration of risk factors.

**Method:** A retrospective study of prevalence has been carried out, in which the medical records of children diagnosed with sensorineural hearing loss born within 2005 and 2012 in the health centers of study have been analyzed.

**Results:** Of the 32 children with sensorineural hearing loss, 16 passed the OAE, 12 did not passed the OAE, and in four they were not carried out. Of the children who passed the OAE, 57% have severe hearing loss. 66% of children with hearing loss presented a risk factor for hearing loss at birth, being the most frequent family history of hearing loss, but only 7% of those with family history of hearing loss were included in the risk group.

**Conclusion:** The results of the study indicate that the late diagnosis of hearing loss is related to the presence of false negatives to the OAE and the non-registration of risk factors.

[Matulat P, Lepper I, Böttcher P, Parfitt R, Oswald H, Am Zehnhoff-Dinnesen A, Deuster D.](#)  
[Two-Way Radio Modem Data Transfer for Newborn Hearing Screening Devices.](#)

[Telemed J E Health.](#) 2016 Jun 6. [Epub ahead of print].

**Introduction:** The success of a newborn hearing screening program depends on successful tracking and follow-up to ensure that children who have had positive screening results in the first few days of life receive appropriate and timely diagnostic and intervention services. The easy availability, through a suitable infrastructure, of the data necessary for the tracking, diagnosis, and care of children concerned is a major key to enhancing the quality and efficiency of newborn hearing screening programs.

**Materials and Methods:** Two systems for the automated two-way transmission of newborn hearing screening and configuration data, based on mobile communication technology, for the screening devices MADSEN AccuScreen® and Natus Echo-Screen® were developed and tested in a field study. Radio modem connections were compared with conventional analogue modem transmissions from Natus Echo-Screen devices for duration, transmission rate, number of lost connections, and frequency of use.

**Results:** The average session duration was significantly lower with the MADSEN AccuScreen (12s) and Natus Echo-Screen both with radio modem (15s) than the Natus Echo-Screen with analogue modem (108s). The transmission rate was significantly higher (898 and 1,758 vs. 181 bytes/s) for the devices with radio modems. Both radio modem devices had significantly lower rates of broken connections after initial connection (2.1 and 0.9 vs. 5.5%). An increase in the frequency of data transmission from the clinics with mobile radio devices was found.

**Conclusion:** The use of mobile communication technology in newborn hearing screening devices offers improvements in the average session duration, transmission rate, and reliability of the connection over analogue solutions. We observed a behavioral change in clinical staff using the new technology: the data exchange with the tracking center is more often used. The requirements for on-site support were reduced. These savings outweigh the small increase in costs for the Internet service provider.

[Mehta D, Noon SE, Schwartz E, Wilkens A, Bedoukian EC, Scarano I, Crenshaw EB 3rd, Krantz ID.](#)

[Outcomes of Evaluation and Testing of 660 Individuals with Hearing Loss in a Pediatric Genetics of Hearing Loss Clinic.](#)

[Am J Med Genet A.](#) 2016 Oct;170(10):2523-30. doi: 10.1002/ajmg.a.37855. Epub 2016 Aug 2.

**Abstract:** Hearing loss is a relatively common condition in children, occurring in approximately 2 out of every 1,000 births with approximately 50% of reported diagnoses having a primary genetic etiology. Given the prevalence and genetic component of hearing loss, coupled with a trend toward early diagnosis with the institution of universal newborn hearing screening, The Genetics of Hearing Loss Clinic was established at The Children's Hospital of Philadelphia to manage the diagnosis, testing, and genetic counseling for individuals and families. This paper described a cohort of 660 individuals with a diagnosis of hearing loss evaluated between July 2008 and July 2015 in the Genetics of Hearing Loss Clinic. To elucidate the cause of hearing loss in this cohort for better management and prognostication, testing included single nucleotide polymorphism chromosomal microarray, hearing loss next generation sequencing panel, and additional clinical tests inclusive of thyroid and renal function studies, temporal bone magnetic resonance imaging, and electrocardiogram. Of those evaluated, most had bilateral sensorineural hearing loss, occurring in 489/660 (74%). Additionally, 612/660 (93%) of patients presented with a nonsyndromic form of hearing loss (no other observed clinical findings at the time of exam), of which pathogenic mutations in GJB2 were most prevalent. Of the individuals with syndromic manifestations (48/660), Usher and Waardenburg syndrome were most commonly observed. A family history of hearing loss (first degree relative) was present in 12.6% of families with available information. Through molecular analyses, clinical examination, and laboratory testing, a definitive etiologic diagnosis was established in 157/660 (23.8%) of individuals. © 2016 Wiley Periodicals, Inc.

[Mena-Domínguez EA, Benito-Orejas JI, Ramírez-Cano B, Morais-Pérez D, Muñoz-Moreno MF.](#)

[High Frequency Tympanometry \(1000Hz\) in Young Infants and its Comparison with Otoacoustic Emissions, Otomicroscopy and 226Hz Tympanometry.](#)

[Acta Otorrinolaringol Esp.](#) 2016 May 12. pii: S0001-6519(16)00037-6. doi: 10.1016/j.otorri.2016.01.001.

**Introduction and Objective:** In the first 6 months of life, 226Hz tympanometry is considered an ineffective procedure for the diagnosis of otitis media with effusion. With the introduction of universal hearing screening, the use of high frequency 1000Hz (1kHz) tympanometry has been recommended. To optimize the diagnosis of neonatal hearing loss, we present this comparison, from the clinical point of view, of the results of 226Hz and 1kHz tympanometry in infants.

**Materials and Methods:** We designed a prospective study of 100 children under 9 months of age proceeding from our hearing screening program. We compare the result of tympanometry with binocular microscopy and transient evoked otoacoustic emissions.

**Results:** The application of transient otoacoustic emissions, otomicroscopy and 226Hz and 1kHz tympanometry has shown its usefulness in the management of otitis media with effusion of young infants, with a similar effectiveness between the 4 tests.

**Conclusion:** The joint use of otomicroscopy, transient otoacoustic emissions and 226Hz and 1kHz tympanometry, has allowed us to diagnose otitis media with effusion in young infants more accurately than each test separately. We recommend initial use of 1kHz tympanometry, at least in children younger than 7 months, but in the presence of hearing loss or an unclear result, 226Hz tympanometry is a good diagnostic complement.

[Naing ZW, Scott GM, Shand A, Hamilton ST, van Zuylen WJ, Basha J, Hall B, Craig ME, Rawlinson WD.](#)

[Congenital Cytomegalovirus Infection in Pregnancy: A Review of Prevalence, Clinical Features, Diagnosis and Prevention.](#)

[Aust N Z J Obstet Gynaecol.](#) 2016 Feb;56(1):9-18. doi: 10.1111/ajo.12408. Epub 2015 Sep 22.

**Abstract:** Human cytomegalovirus (CMV) is under-recognised, despite being the leading infectious cause of congenital malformation, affecting ~0.3% of Australian live births. Approximately 11% of infants born with congenital CMV infection are symptomatic, resulting in clinical manifestations, including jaundice, hepatosplenomegaly, petechiae, microcephaly, intrauterine growth restriction and death. Congenital CMV infection may cause severe long-term sequelae, including progressive sensorineural hearing loss and developmental delay in 40-58% of symptomatic neonates, and ~14% of initially asymptomatic infected neonates. Up to 50% of maternal CMV infections have nonspecific clinical manifestations, and most remain undetected unless specific serological testing is undertaken. The combination of serology tests for CMV-specific IgM, IgG and IgG avidity provide improved distinction between primary and secondary maternal infections. In pregnancies with confirmed primary maternal CMV infection, amniocentesis with CMV-PCR performed on amniotic fluid, undertaken after 21-22 weeks gestation, may determine whether maternofetal virus transmission has occurred. Ultrasound and, to a lesser extent, magnetic resonance imaging are valuable tools to assess fetal structural and growth abnormalities, although the absence of fetal abnormalities does not exclude fetal damage. Diagnosis of congenital CMV infection at birth or in the first 3 weeks of an infant's life is crucial, as this should prompt interventions for prevention of delayed-onset hearing loss and neurodevelopmental delay in affected infants. Prevention strategies should also target mothers because increased awareness and hygiene measures may reduce maternal infection. Recognition of the importance of CMV in pregnancy and in neonates is increasingly needed, particularly as therapeutic and preventive interventions expand for this serious problem.

[Nittrouer S.](#)

### [Beyond Early Intervention: Supporting Children With CIs Through Elementary School.](#)

*Otol Neurotol.* 2016 Feb;37(2):e43-9. doi: 10.1097/MAO.0000000000000906.

**Background:** The development of cochlear implants (CIs) and the broader availability of early intervention, made possible by newborn hearing screening, have raised prospects that deaf children can be mainstreamed at the start of elementary school and fare well with minimal support. This report examines the veracity of that perspective.

**Methods:** This report specifically: (1) reviews progress made by deaf children in spoken language acquisition over the past 25 years; (2) presents data collected from 104 children in the early elementary grades (49 with normal hearing (NH) and 55 with severe-to-profound hearing loss who use CIs); (3) describes language acquisition that typically occurs in elementary school; and (4) highlights intervention strategies for school-age deaf children with CIs.

**Results:** The spoken language skills of deaf children have improved thanks to CIs and early intervention, but remain below those of children with NH. The amount of deficit varies across the language construct examined, with the greatest deficit found for skills dependent upon phonological (speech-sound) sensitivity, and the mildest associated with morphosyntactic (grammatical) skills. There is substantial development in both phonological and morphosyntactic skills that typically occurs during the elementary school years.

**Conclusion:** Both the data and theoretical models of language acquisition indicate that even with the availability of CIs and early intervention, deaf children are behind their peers with NH when they enter school. And there is much language learning that lies ahead for them. Thus, there is a need for us to enhance.

[Nittrouer S.](#), [Lowenstein JH.](#), [Holloman C.](#)

### [Early Predictors of Phonological and Morphosyntactic Skills in Second Graders with Cochlear Implants.](#)

*Res Dev Disabil.* 2016 Aug;55:143-60. doi: 10.1016/j.ridd.2016.03.020. Epub 2016 Apr 12.

**Purpose:** Newborn hearing screening has made it possible to provide early treatment of hearing loss to more children than ever before, raising expectations these children will be able to attend regular schools. But continuing deficits in spoken language skills have led to challenges in meeting those expectations. This study was conducted to (1) examine two kinds of language skills (phonological and morphosyntactic) at school age (second grade) for children with cochlear implants (CIs); (2) see which measures from earlier in life best predicted performance at second grade; (3) explore how well these skills supported other cognitive and language functions; and (4) examine how treatment factors affected measured outcomes.

**Methods:** Data were analyzed from 100 second-grade, monolingual English-speaking children: 51 with CIs and 49 with normal hearing (NH). Ten measures of spoken language and related functions were collected: three each of phonological and morphosyntactic skills; and four of other cognitive and language functions. Six measures from preschool and seven from kindergarten served as predictor variables. The effects of treatment variables were examined.

**Results:** Children with CIs were more delayed acquiring phonological than morphosyntactic skills. Mean length of utterance at earlier ages was the most consistent predictor of both phonological and morphosyntactic skills at second grade. Early bimodal stimulation had a weak, but positive effect on phonological skills at second grade; sign language experience during preschool had a negative effect on morphosyntactic structures in spoken language.

**Conclusion:** Children with CIs are delayed in language acquisition, and especially so in phonological skills. Appropriate testing and treatments can help ameliorate these delays.

[Noble JH.](#), [Hedley-Williams AJ.](#), [Sunderhaus L.](#), [Dawant BM.](#), [Labadie RF.](#), [Camarata SM.](#), [Gifford RH.](#)

### [Initial Results With Image-guided Cochlear Implant Programming in Children.](#)

*Otol Neurotol.* 2016 Feb;37(2):e63-9. doi: 10.1097/MAO.0000000000000909.

**Hypothesis:** Image-guided cochlear implant (CI) programming can improve hearing outcomes for pediatric CI recipients.

**Background:** CIs have been highly successful for children with severe-to-profound hearing loss, offering potential for mainstreamed education and auditory-oral communication. Despite this, a significant number of recipients still experience poor speech understanding, language delay, and, even among the best performers, restoration to normal auditory fidelity is rare. Although significant research efforts have been devoted to improving stimulation strategies, few developments have led to significant hearing improvement over the past two decades. Recently introduced techniques for image-guided CI programming (IGCIP) permit creating patient-customized CI programs by making it possible, for the first time, to estimate the position of implanted CI electrodes relative to the nerves they stimulate using CT images. This approach permits identification of electrodes with high levels of stimulation overlap and to deactivate them from a patient's map. Previous studies have shown that IGCIP can significantly improve hearing outcomes for adults with CIs.

**Methods:** The IGCIP technique was tested for 21 ears of 18 pediatric CI recipients. Participants had long-term experience with their CI (5 mo to 13 yr) and ranged in age from 5 to 17 years old. Speech understanding was assessed after approximately 4 weeks of experience with the IGCIP map.

**Results:** Using a two-tailed Wilcoxon signed-rank test, statistically significant improvement ( $p < 0.05$ ) was observed for word and sentence recognition in quiet and noise, as well as pediatric self-reported quality-of-life (QOL) measures.

**Conclusion:** Our results indicate that image guidance significantly improves hearing and QOL outcomes for pediatric CI recipients.

[Núñez-Batalla F.](#), [Jáudenes-Casabón C.](#), [Sequí-Canet JM.](#), [Vivanco-Allende A.](#), [Zubicaray-Ugarteche J.](#)

[Cabanillas-Farpón R.](#)

### [Aetiological Diagnosis of Child Deafness: CODEPEH Recommendations.](#)

*Acta Otorrinolaringol Esp.* 2016 Sep 16. pii: S0001-6519(16)30050-4. doi: 10.1016/j.otorri.2016.05.002.

**Abstract:** Important progress in the fields of molecular genetics (principally) and diagnostic imaging, together with the lack of a consensus protocol for guiding the diagnostic process after confirming deafness by neonatal screening, have led to this new work document drafted by the Spanish Commission for the Early Detection of Child Deafness (Spanish acronym: CODEPEH). This 2015 Recommendations Document, which is based on the most recent scientific evidence, provides guidance to professionals to support them in making decisions regarding aetiological diagnosis. Such diagnosis should be performed without delay and without impeding early intervention. Early identification of the causes of deafness offers many advantages: it prevents unnecessary trouble for the families, reduces health system expenses caused by performing different tests, and provides prognostic information that may guide therapeutic actions.

[Núñez-Batalla F, Noriega-Iglesias S, Guntín-García M, Carro-Fernández P, Llorente-Pendás JL.](#)

### [Auditory-Steady-State Response Reliability in the Audiological Diagnosis After Neonatal Hearing Screening.](#)

[Acta Otorrinolaringol Esp.](#) 2016 Jul-Aug;67(4):193-200. doi: 10.1016/j.otorri.2015.06.003. Epub 2015 Oct 9.

**Introduction and Objectives:** Conventional audiometry is the gold standard for quantifying and describing hearing loss. Alternative methods become necessary to assess subjects who are too young to respond reliably. Auditory evoked potentials constitute the most widely used method for determining hearing thresholds objectively; however, this stimulus is not frequency specific. The advent of the auditory steady-state response (ASSR) leads to more specific threshold determination. The current study describes and compares ASSR, auditory brainstem response (ABR) and conventional behavioural tone audiometry thresholds in a group of infants with various degrees of hearing loss.

**Methods:** A comparison was made between ASSR, ABR and behavioural hearing thresholds in 35 infants detected in the neonatal hearing screening program.

**Results:** Mean difference scores ( $\pm$ SD) between ABR and high frequency ABR thresholds were 11.2 dB ( $\pm$ 13) and 10.2 dB ( $\pm$ 11). Pearson correlations between the ASSR and audiometry thresholds were 0.80 and 0.91 (500Hz); 0.84 and 0.82 (1000Hz); 0.85 and 0.84 (2000Hz); and 0.83 and 0.82 (4000Hz).

**Conclusion:** The ASSR technique is a valuable extension of the clinical test battery for hearing-impaired children.

[Núñez-Batalla F, Jáudenes-Casaubón C, Sequí-Canet JM, Vivanco-Allende A, Zubicaray-Ugarteche J.](#)

### [2014 CODEPEH Recommendations: Early Detection of Late Onset Deafness, Audiological Diagnosis, Hearing Aid Fitting and Early Intervention.](#)

[Acta Otorrinolaringol Esp.](#) 2016 Jan-Feb;67(1):45-53. doi: 10.1016/j.otorri.2015.05.007. Epub 2015 Oct 9.

**Abstract:** The latest scientific literature considers early diagnosis of deafness as the key element to define the educational and inclusive prognosis of the deaf child, because it allows taking advantage of the critical period of development (0-4 years). Highly significant differences exist between deaf people who have been stimulated early and those who have received late or improper intervention. Early identification of late-onset disorders requires special attention and knowledge on the part of every childcare professional. Programs and additional actions beyond neonatal screening should be designed and planned to ensure that every child with a significant hearing loss is detected early. For this purpose, the CODEPEH would like to highlight the need for continuous monitoring of children's auditory health. Consequently, CODEPEH has drafted the recommendations included in the present document.

[Palmer SB, Bednarz SE, Dilaj KA, McDonald AM.](#)

### [Universal Newborn Hearing Screening in Midwifery Education: A Survey.](#)

[J Midwifery Womens Health.](#) 2016 Jul;61(4):435-41. doi: 10.1111/jmwh.12438. Epub 2016 May 10.

**Introduction:** Universal newborn hearing screening has been adopted by all 50 states in the United States. However, there is currently a lack of knowledge about how health care providers learn about universal newborn hearing screening during their education programs. The purpose of this study was to identify whether midwifery education programs in the United States currently include information regarding universal newborn hearing screening in the standard curricula and, if so, what specific information is covered.

**Methods:** A survey that assessed whether specific topics related to universal newborn hearing screening are presented during midwifery education was sent to directors of midwifery education programs.

**Results:** Seventy-one midwifery education program directors were contacted, and the response rate was 38% (27 surveys). Most respondents reported that universal newborn hearing screening is discussed in the program, with the amount of time spent covering these topics varying considerably. Programs provide information about the midwife's role in universal newborn hearing screening, legal obligation to provide hearing screening information, and tests used to complete universal newborn hearing screening. How to complete the hearing screening, counseling for parents about results, and follow-up after a newborn does not pass the screening are topics that were not often discussed. There was no influence of program type or program length on the universal newborn hearing screening content discussed.

**Conclusion:** The majority of midwifery education program directors that responded indicated that their programs include information about universal newborn hearing screening to midwifery students. There is a need for further information and resources specific to universal newborn hearing screening. Providing additional information to midwifery students about newborn hearing screening may result in increased awareness and education for families.

[Peng Q, Huang S, Liang Y, Ma K, Li S, Yang L, Li W, Ma Q, Liu Q, Zhong B, Lu X.](#)

### [Concurrent Genetic and Standard Screening for Hearing Impairment in 9317 Southern Chinese Newborns.](#)

[Genet Test Mol Biomarkers.](#) 2016 Oct;20(10):603-608. Epub 2016 Aug 19.

**Objective:** The goal of this study was to investigate the use of concurrent genetic screening together with standard newborn hearing screening (NHS) in an effort to provide a scientific basis for the beneficial use of concurrent genetic hearing screening in newborns. Our aim was to improve the neonatal detection rate of hearing impairment and the potential for hearing loss, allowing for increased early intervention and potentially allowing for prevention of later onset hearing loss. This information could also be used to increase the effectiveness of genetic counseling regarding hearing impairment.

**Methods:** A total of 9317 neonates from Children's Hospital of Dongguan and Dongguan People's Hospital were included in this study between January 2015 and October 2015. Twenty hotspot hearing-associated mutations of four common deafness-susceptibility genes (GJB2, GJB3, SLC26A4, and MTRNR1) were analyzed by matrix-assisted laser desorption-ionization time-of-flight mass spectrometry (MALDI-TOF-MS). The results of genetic screening and NHS were concurrently analyzed.

**Results:** A total of 129 infants (1.38%) exhibited hearing loss as determined by otoacoustic emission (OAE) testing. The genetic screening revealed that 348 (3.74%) individuals had at least one mutant allele. In total, 34 (0.36%) of the neonates carried a causal complement of mutations. The overwhelming majority of the genetically referred newborns passed the OAE hearing screening, but could be at risk for later hearing loss.

**Conclusion:** This study furthers the understanding of the etiology of hearing loss and proves that it is beneficial to use genetic screening along with OAE screening of neonates to improve detection rates of at-risk infants. Our results show that this concurrent testing allows for better early identification of infants at risk for hearing loss, which may occur before speech and language development. Prevention of hearing loss can be achieved by avoiding the use of antibiotics containing amino glycosides in infants whose mutations make them extremely sensitive to these antibiotics. This information is also useful in genetic counseling, providing region-specific mutation information.

[Phelan E, Pal R, Henderson L, Green KM, Bruce IA.](#)

### [The Management of Children with Down Syndrome and Profound Hearing Loss.](#)

[Cochlear Implants Int.](#) 2016;17(1):52-7. doi: 10.1179/1754762815Y.0000000019. Epub 2015 Aug 7.

**Introduction:** Although, the association between Down syndrome (DS) and conductive hearing loss is well recognized, the fact that a small proportion of these children may have a severe to profound sensorineural hearing loss that could benefit from cochlear implantation (CI) is less well understood. The management of significant co-morbidities in children with DS can delay initial diagnosis of hearing impairment and assessment of suitability for CI can likewise be challenging, due to difficulties conditioning to behavioural hearing tests.

**Methods:** We performed a retrospective case note review of three children with DS referred to the Manchester Cochlear Implant Programme.

**Results:** Three illustrative cases are described including CI in a 4 years old. Using conventional outcome measurement instruments, the outcome could be considered to be suboptimal with a Categories of Auditory Performance score of 4 at 6 months post-op and at last follow up. In part, this is likely to reflect the delay in implantation, but the role of cognitive impairment must be considered. The cases described emphasize the importance of comprehensive radiological and audiological assessment in children with DS being considered for CI.

**Conclusion:** The influence of cognitive impairment upon outcome of CI must be taken into account, but should not be considered a contra-indication to implantation in children with DS. Benefit that might be considered limited when quantified using existing general outcome measurement instruments, may have a significant impact upon psychosocial development and quality of life in children with significant cognitive impairment, or other additional needs.

[Pimperton H, Blythe H, Kreppner J, Mahon M, Peacock JL, Stevenson J, Terleksi E, Worsfold S, Yuen HM, Kennedy CR.](#)

### [The Impact of Universal Newborn Hearing Screening on Long-Term Literacy Outcomes: A Prospective Cohort Study.](#)

[Arch Dis Child.](#) 2016 Jan;101(1):9-15. doi: 10.1136/archdischild-2014-307516. Epub 2014 Nov 25.

**Objective:** To determine whether the benefits of universal newborn hearing screening (UNHS) seen at age 8 years persist through the second decade.

**Design:** Prospective cohort study of a population sample of children with permanent childhood hearing impairment (PCHI) followed up for 17 years since birth in periods with (or without) UNHS.

**Setting:** Birth cohort of 100 000 in southern England.

**Participants:** 114 teenagers aged 13-19 years, 76 with PCHI and 38 with normal hearing. All had previously their reading assessed aged 6-10 years.

**Interventions:** Birth in periods with and without UNHS; confirmation of PCHI before and after age 9 months.

**Main Outcome Measure:** Reading comprehension ability. Regression modelling took account of severity of hearing loss, non-verbal ability, maternal education and main language.

**Results:** Confirmation of PCHI by age 9 months was associated with significantly higher mean z-scores for reading comprehension (adjusted mean difference 1.17, 95% CI 0.36 to 1.97) although birth during periods with UNHS was not (adjusted mean difference 0.15, 95% CI -0.75 to 1.06). The gap between the reading comprehension z-scores of teenagers with early compared with late confirmed PCHI had widened at an adjusted mean rate of 0.06 per year (95% CI -0.02 to 0.13) during the 9.2-year mean interval since the previous assessment.

**Conclusion:** The benefit to reading comprehension of confirmation of PCHI by age 9 months increases during the teenage years. This strengthens the case for UNHS programmes that lead to early confirmation of permanent hearing loss.

[Pitaro J, Al Masaoudi L, Motallebzadeh H, Funnell WR, Daniel SJ.](#)

### [Wideband Reflectance Measurements in Newborns: Relationship to Otoscopic Findings.](#)

[Int J Pediatr Otorhinolaryngol.](#) 2016 Jul;86:156-60. doi: 10.1016/j.ijporl.2016.04.036. Epub 2016 May 2.

**Objectives:** Newborn hearing screening includes testing with otoacoustic emissions and the auditory brainstem response. Unfortunately, both tests are affected by the presence of material in the ear canal and middle ear such as vernix, meconium, and amniotic fluid. The objective of this study was to determine to what extent occlusion of the ear canal as seen on otoscopy affects wideband energy reflectance measurements in newborns. A secondary objective was to obtain additional normative wideband reflectance data in newborns.

**Methods:** Newborns from a well-baby nursery were enrolled. Wideband energy reflectance measurements and otoscopy were done immediately after the hearing screening. Occlusion of the ear canal as seen on otoscopy was described on a scale of 0-100%.

**Results:** A total of 156 babies were enrolled (mean age = 25 hours). A statistically significant difference in the reflectance at ambient pressure was found between the 0-70% and 80-100% occlusion groups. There was no significant difference in reflectance between the right and the left ears. The median reflectance pattern generally followed that of previous studies but in certain frequency regions the present reflectance values were higher.

**Conclusion:** A significant increase in reflectance occurs when 70%-80% of the ear-canal diameter is occluded. Taking otoscopy findings into account may improve the interpretation of reflectance measurements. However, further studies are required to better establish the relationship between canal occlusion and reflectance.

[Poonual W, Navacharoen N, Kongsanarak J, Namwongprom S.](#)

### [Risk Factors for Hearing Loss in Infants Under Universal Hearing Screening Program in Northern Thailand.](#)

[J Multidiscip Healthc.](#) 2015 Dec 24;9:1-5. doi: 10.2147/JMDH.S92818. eCollection 2016.

**Objectives:** To define the risk factors for hearing loss in infants (aged 3 months) under universal hearing screening program.

**Materials and Methods:** A total of 3,120 infants (aged 3 months) who underwent hearing screening using a universal hearing screening program using automated otoacoustic emission test between November 1, 2010 and May 31, 2012 in Uttaradit Hospital, Buddhachinaraj Hospital, and Sawanpracharuk Hospital (tertiary hospitals) located in Northern Thailand were included in this prospective cohort study.

**Results:** Of the 3,120 infants, 135 (4.3%) were confirmed to have hearing loss with the conventional otoacoustic emission test. Five of these 135 infants (3.7%) with hearing loss showed test results consistent with auditory brainstem responses. From the univariable analysis, there were eleven potential risk factors associated with hearing deterioration. On multivariable analysis, the risk factors independently associated with hearing loss at 3 months were birth weight 1,500-2,500 g (risk ratio [RR] 1.6, 95% confidence interval [CI] 1.1-2.6), APGAR score <6 at 5 minutes (RR 2.2, 95% CI 1.1-4.4), craniofacial anomalies (RR 2.5, 95% CI 1.6-4.2), sepsis (RR 1.8, 95% CI 1.0-3.2), and ototoxic exposure (RR 4.1, 95% CI 1.9-8.6).

**Conclusion:** This study concluded that low birth weight, APGAR score <6 at 5 minutes, craniofacial anomalies, sepsis, and ototoxic exposure are the risk factors for bilateral hearing loss in infants (aged 3 months) and proper tests should be performed to identify these risk factors. As an outcome, under the present circumstances, it is suggested that infirmary/physicians/general practitioners/health action centers/polyclinics should carry out universal hearing screening in all infants before 36 weeks. The public health policy of Thailand regarding a universal hearing screening program is important for the prevention of disability and to enhance people's quality of life.

[Psarros C, Love S.](#)

### [The Role of the World Health Organization's International Classification of Functioning, Health and Disability in Models of Infant Cochlear Implant Management.](#)

[Semin Hear. 2016 Aug;37\(3\):272-90. doi: 10.1055/s-0036-1584414.](#)

**Abstract:** Newborn hearing screening has led to the early diagnosis of hearing loss in neonates and early device fitting is common, based primarily on electrophysiologic and radiologic information, with some supplementary behavioral measures. Such early fitting of hearing devices, in particular cochlear implants (CIs), has been beneficial to the majority of children implanted under the age of 12 months who meet the cochlear implant candidacy criteria. Comorbidities are common in children with hearing loss, although they may not be evident in neonates and may not emerge until later in infants. Evidence suggests that the child's outcomes are strongly influenced by a range of environmental factors including emotional and social support from the immediate and extended family. Consequently, such factors are important in service planning and service delivery for babies and children receiving CIs. The World Health Organization's International Classification of Functioning, Health and Disability (ICF) can provide a framework to facilitate the holistic management of pediatric cochlear implant recipients. The ICF also can be used to map the progress of recipients over time to highlight emerging issues that require intervention. This article will discuss our preliminary use of the ICF to establish clinical practice; develop advocacy skills among clients and their families; identify eligibility for services such as support in educational settings; enable access to modes of service delivery such as telepractice; provide a conceptual framework for policy and program development for pediatric cochlear implant recipients (i.e., in both disability and health services); and, most importantly, establish a clear pathway for the longitudinal management of the cochlear implant in a child's future. It is anticipated that this model will be applied to other populations receiving cochlear implants through our program.

[Pynnonen MA, Handelsman JA, King EF, Singer DC, Davis MM, Lesperance MM.](#)

### [Parent Perception of Newborn Hearing Screening: Results of a US National Survey.](#)

[JAMA Otolaryngol Head Neck Surg. 2016 Jun 1;142\(6\):538-43. doi: 10.1001/jamaoto.2015.3948.](#)

**Importance:** An unacceptably high number of children who do not pass universal newborn hearing screening (UNHS) are lost to follow-up.

**Objectives:** To provide insight into parent recall of UNHS.

**Design, Setting, and Participants:** In this nationally representative cross-sectional survey, 2144 US parent households were surveyed in May 2012 using the Knowledge Panel. Responses of parents whose children were born before vs after UNHS implementation were compared.

**Main Outcomes and Measures:** Outcome measures included recall of hearing screen at birth, hearing screen results, and recommendations for follow-up. All outcome measures were based on parent recall and report. Descriptive statistics and multiple logistic regression analyses were used.

**Results:** The study participants included 1539 parent households and 605 nonparent households. Of the 1539 parent households surveyed, the mean age of the parents was 38.8 years (range, 18-88 years), the mean age of the children was 10.2 years (range, 0-17 years), and the mean age of children with hearing loss was 12.1 years (range, 0-17 years). A total of 1539 parents (55.8%) were women. Only 62.9% of parents (unweighted n = 950) recalled a newborn hearing screen, and among those children with risk indicators for hearing loss (n = 587), only 68.6% (unweighted n = 385) recalled a hearing screen. Higher parent educational level (odds ratio [OR], 2.27; 95% CI, 1.17-4.41, for some college and OR, 2.41; 95% CI, 1.22-4.78, for a bachelor's degree; P = .03), younger age of the child (OR, 1.16; 95% CI, 1.11-1.23; P < .001), and the presence of any risk indicator for hearing loss (OR, 1.5; 95% CI, 1.13-2.13; P = .007) were associated with parent recall of hearing screen. Reported pass rates were higher than expected. Parent recall of follow-up recommendations was not always consistent with guidelines.

**Conclusion:** Although this study is inherently limited by recall bias, the findings indicate a lack of parent awareness of UNHS. Changes in the system of reporting UNHS results are necessary to improve parent recall of screen results and improve follow-up for children who do not pass the screen.

[Raine C, Atkinson H, Strachan DR, Martin JM.](#)

### [Access to Cochlear Implants: Time to reflect.](#)

[Cochlear Implants Int. 2016 Apr;17 Suppl 1:42-6. doi: 10.1080/14670100.2016.1155808.](#)

**Abstract:** Cochlear implant (CI) intervention is expensive and accessed mainly by developed countries. The introduction of Universal Newborn Hearing Screening and funding via a public health service give children better access to CIs. However, for adults large disparities exist between utilization and estimated prevalence. In the UK CI selection criteria are restrictive compared with many other countries. Improved audiological awareness and screening programmes for adults would improve access to hearing technologies that would improve health and quality of life. Hearing loss itself has significant medical and financial burdens on society and by investing in early intervention and using best technology this would mitigate some of the rising associated medical costs.

[Raveh E, Ulanovski D, Attias J, Shkedy Y, Sokolov M.](#)

### [Acute Mastoiditis in Children with a Cochlear Implant.](#)

[Int J Pediatr Otorhinolaryngol. 2016 Feb;81:80-3. doi: 10.1016/j.ijporl.2015.12.016. Epub 2015 Dec 31.](#)

**Objectives:** Cochlear implantation is performed at a young age, when children are prone to acute otitis media. Acute mastoiditis is the most common complication of otitis media, but data on its management in the presence of a cochlear implant are sparse. The objective of this study was to assess the characteristics, treatment, and outcome of acute mastoiditis in children with a cochlear implant.

**Methods:** The medical files of all children who underwent cochlear implantation at a pediatric tertiary medical center in 2000-2014 were retrospectively reviewed. Those diagnosed with acute mastoiditis after implantation were identified, and data were collected on demographics, history, presentation, method of treatment, complications, association with untreated otitis media with effusion, and long-term middle-ear sequelae.

**Results:** Of the 370 children (490 ears) who underwent cochlear implantation, 13 (3.5%) were treated for acute mastoiditis (median age at acute mastoiditis, 32 months). Nine had a pre-implantation history of chronic secretory or acute recurrent otitis media, and 5 had been previously treated with ventilation tubes. In all 9 children who had unilateral cochlear implant, the acute mastoiditis episode occurred in the implanted ear. The time from implantation to mastoiditis was 5-61 months. The same treatment protocol as for normal-hearing children was followed, with special attention to the risk of central nervous system complications. Primary treatment consisted of myringotomy with intravenous administration of wide-spectrum antibiotics. Surgical drainage was performed in 8 out of 13 patients, with (n=7) or without (n=1) ventilation-tube insertion, to treat subperiosteal abscess or because of lack of symptomatic improvement. There were no cases of intracranial complications or implant involvement or need for a wider surgical approach. No middle-ear pathology was documented during the average 3.8-year follow-up.

**Conclusion:** The relatively high rate of acute mastoiditis and subperiosteally abscess in children with a cochlear implant, predominantly involving the implanted ear, supports the suggestion that recent mastoiditis may be a risk factor for these complications. Despite the frequent need for drainage, more extensive surgery is usually unnecessary, and recovery is complete and rapid. As infections can occur even years after cochlear implantation, children with otitis media should be closely followed, with possible re-introduction of ventilation tubes.

[Ravi R, Gunjawate DR, Yerraguntla K, Lewis LE, Driscoll C, Rajashekhar B.](#)

### [Follow-up in newborn hearing screening - A systematic review.](#)

*Int J Pediatr Otorhinolaryngol.* 2016 Nov;90:29-36. doi: 10.1016/j.ijporl.2016.08.016. Epub 2016 Aug 26.

**Introduction:** The quality and efficiency of newborn hearing screening programs (NHS) rely heavily on appropriate follow-up. The Joint Committee on Infant Hearing recommends a follow-up rate of more than 95% of infants who fail the initial hearing screening. However, a 70% benchmark is considered to be more feasible. This high loss to follow-up (LTF) rate acts as a threat to the overall success of NHS programs. The objective of the study was to identify and examine the reported rates of LTF, attributed reasons for LTF and strategies undertaken to reduce LTF.

**Methods:** Using a systematic search, articles published between 2005 to December 2015 were identified from PubMed/Medline, Cumulative Index to Nursing and Allied Health Literature (CINAHL), Educational Resources Information Center (ERIC), Scopus, Ovid, ProQuest, and Cochrane Library. To be included in the review, the study should be exploring the loss to follow-up or drop-out rate in newborn hearing screening programs and be published in an indexed peer-reviewed journal in the English language. The main outcome measures were overall rate of LTF, factors leading to LTF and measures adopted to overcome LTF.

**Results:** 53 articles were short-listed for data extraction. Out of these, 27 were single-centre studies, 19 were multi-centre, 3 compared multiple databases, and 4 used survey-based methods. Overall LTF rates of 20% in single-centre and 21% in multiple-centre studies were observed. Educational disparity and lack of adequate knowledge among parents were associated with LTF. The most commonly used strategy to overcome LTF suggested by studies was the use of an adequate data management system.

**Conclusion:** This review is a novel attempt to explore the LTF among NHS studies, reasons for LTF and strategies to reduce LTF. This review can act as a basis for planning and execution of effective NHS programs.

[Ravi R, Yerraguntla K, Gunjawate DR, Rajashekhar B, Lewis LE, Guddattu V.](#)

### [Knowledge and Attitude \(KA\) Survey Regarding Infant Hearing Loss in Karnataka, India.](#)

*Int J Pediatr Otorhinolaryngol.* 2016 Jun;85:1-4. doi: 10.1016/j.ijporl.2016.03.012. Epub 2016 Mar 23.

**Introduction:** The support provided and the decisions taken by mothers determine the success of Universal Newborn Hearing Screening (UNHS). Attempts at exploring the existing knowledge-attitude among mothers is crucial to create/modify the existing screening programs. The present study attempts to explore the knowledge and attitude toward infant hearing loss (HL) among mothers of newborns in the Indian state of Karnataka.

**Methods:** A cross-sectional survey was conducted among 219 mothers of newborns in Karnataka, India. The questionnaire was framed from existing literature and consisted of 19 questions assessing knowledge and attitude toward infant HL to be rated on a three-point scale (no, not sure, yes). Descriptive statistics and Cronbach's  $\alpha$  were used to analyze the data.

**Results:** Mothers exhibited good knowledge of risk factors; noise (70.3%) and ear discharge (54.3%). More than 75% agreed that treatment for HL is available and that these children can attend school. The questions of superstitions and cultural beliefs yielded mixed responses. A large number of mothers expressed desire to have their children tested at birth (84.9%) and were concerned about their children's hearing (87.7%). Yet only 54.3% stated that they would allow their children to wear hearing aids.

**Summary and Conclusion:** The present study is an attempt to understand the knowledge and attitude of mothers toward infant HL in Karnataka and facilitate identification of potential areas of less knowledge as a reference for endeavors of enhancement. It further highlights the need for implementing public awareness programs to improve knowledge and attitude of mothers toward infant HL for better implementation of UNHS.

[Ravi R, Gunjawate DR, Yerraguntla K, Rajashekhar B, Lewis LE.](#)

### [Knowledge and Attitude of Parents/Caregivers Towards Hearing Loss and Screening in Newborns - A Systematic Review.](#)

*Int J Audiol.* 2016 Dec;55(12):715-722. Epub 2016 Aug 15.

**Objective:** The parents/caregivers of a newborn play a pivotal role in the process of hearing screening and intervention. The decisions taken by them depend on their knowledge and attitude. The purpose of this study was to review the literature systematically on knowledge and attitude of parents/caregivers towards infant hearing loss and newborn hearing screening.

**Design:** A systematic search was conducted using electronic databases for the periods from 1990 to March 2016. Two authors scrutinized the studies and extracted the data based on predetermined criteria.

**Study Sample:** Ten studies.

**Results:** Ear discharge was correctly identified as a risk factor for hearing loss along with measles, drugs/medication, family history, congenital causes and noise exposure. The studies revealed mixed results for knowledge about newborn hearing screening. Overall, the parents/caregivers showed positive attitudes towards hearing screening and intervention options. However, due to heterogeneity in the studies, it's hard to derive a conclusion.

**Summary and Conclusion:** The present review sheds light on the common areas of misconception among parents/caregivers about risk factors of infant hearing loss and newborn hearing screening. The review also draws attention to the need to have more studies exploring this knowledge and attitude of parents/caregivers among diverse populations.

[Ribeiro GE, Silva DP, Montovani JC.](#)

### [Transient Evoked Otoacoustic Emissions and Auditory Brainstem Response in Infants with Perinatal Asphyxia.](#)

*Int J Pediatr Otorhinolaryngol.* 2016 Oct;89:136-9. doi: 10.1016/j.ijporl.2016.08.009. Epub 2016 Aug 15.

**Objective:** The objective of this study was to verify the effects of perinatal asphyxia on different parts of the auditory system.

**Methods:** This was a non-concurrent cohort study conducted on a fixed population in a tertiary public hospital. Participants included 181 infants born at term who underwent the transient evoked otoacoustic emission test as a part of a neonatal hearing screening program, with a "pass" result in both ears, and by auditory brainstem response testing. The infants were divided into 3 groups: G1, 20 infants who had perinatal asphyxia; G2, 111 infants with an Apgar score lower than 4 in the first minute and/or lower than 6 in the fifth minute (called "low Apgar" at birth); and G3, 50 infants with first- and fifth-minute Apgar scores  $\geq 7$ .

**Results:** The signal-to-noise ratio of transient evoked otoacoustic emissions were greater in G3 compared with G1 and G2 at 4 kHz frequency for males. An increased latency of waves I and III in the auditory brainstem response of male infants in G1 was observed.

**Conclusion:** This study demonstrated that alterations occurred in both the cochlear and the neural components in male infants who had perinatal asphyxia.

[Rosenfeld RM](#), [Shin JJ](#), [Schwartz SR](#), [Coggins R](#), [Gagnon L](#), [Hackell JM](#), [Hoelting D](#), [Hunter LL](#), [Kummer AW](#), [Payne SC](#), [Poe DS](#), [Veling M](#), [Vila PM](#), [Walsh SA](#), [Corrigan MD](#).

### **Clinical Practice Guideline: Otitis Media with Effusion (Update).**

*Otolaryngol Head Neck Surg.* 2016 Feb;154(1 Suppl):S1-S41. doi: 10.1177/0194599815623467.

**Objective:** This update of a 2004 guideline codeveloped by the American Academy of Otolaryngology-Head and Neck Surgery Foundation, the American Academy of Pediatrics, and the American Academy of Family Physicians, provides evidence-based recommendations to manage otitis media with effusion (OME), defined as the presence of fluid in the middle ear without signs or symptoms of acute ear infection. Changes from the prior guideline include consumer advocates added to the update group, evidence from 4 new clinical practice guidelines, 20 new systematic reviews, and 49 randomized control trials, enhanced emphasis on patient education and shared decision making, a new algorithm to clarify action statement relationships, and new and expanded recommendations for the diagnosis and management of OME.

**Purpose:** The purpose of this multidisciplinary guideline is to identify quality improvement opportunities in managing OME and to create explicit and actionable recommendations to implement these opportunities in clinical practice. Specifically, the goals are to improve diagnostic accuracy, identify children who are most susceptible to developmental sequelae from OME, and educate clinicians and patients regarding the favorable natural history of most OME and the clinical benefits for medical therapy (eg, steroids, antihistamines, decongestants). Additional goals relate to OME surveillance, hearing and language evaluation, and management of OME detected by newborn screening. The target patient for the guideline is a child aged 2 months through 12 years with OME, with or without developmental disabilities or underlying conditions that predispose to OME and its sequelae. The guideline is intended for all clinicians who are likely to diagnose and manage children with OME, and it applies to any setting in which OME would be identified, monitored, or managed. This guideline, however, does not apply to patients <2 months or >12 years old.

**Action Statements:** The update group made strong recommendations that clinicians (1) should document the presence of middle ear effusion with pneumatic otoscopy when diagnosing OME in a child; (2) should perform pneumatic otoscopy to assess for OME in a child with otalgia, hearing loss, or both; (3) should obtain tympanometry in children with suspected OME for whom the diagnosis is uncertain after performing (or attempting) pneumatic otoscopy; (4) should manage the child with OME who is not at risk with watchful waiting for 3 months from the date of effusion onset (if known) or 3 months from the date of diagnosis (if onset is unknown); (5) should recommend against using intranasal or systemic steroids for treating OME; (6) should recommend against using systemic antibiotics for treating OME; and (7) should recommend against using antihistamines, decongestants, or both for treating OME. The update group made recommendations that clinicians (1) should document in the medical record counseling of parents of infants with OME who fail a newborn screening regarding the importance of follow-up to ensure that hearing is normal when OME resolves and to exclude an underlying sensorineural hearing loss; (2) should determine if a child with OME is at increased risk for speech, language, or learning problems from middle ear effusion because of baseline sensory, physical, cognitive, or behavioral factors; (3) should evaluate at-risk children for OME at the time of diagnosis of an at-risk condition and at 12 to 18 months of age (if diagnosed as being at risk prior to this time); (4) should not routinely screen children for OME who are not at risk and do not have symptoms that may be attributable to OME, such as hearing difficulties, balance (vestibular) problems, poor school performance, behavioral problems, or ear discomfort; (5) should educate children with OME and their families regarding the natural history of OME, need for follow-up, and the possible sequelae; (6) should obtain an age-appropriate hearing test if OME persists for 3 months or longer OR for OME of any duration in an at-risk child; (7) should counsel families of children with bilateral OME and documented hearing loss about the potential impact on speech and language development; (8) should reevaluate, at 3- to 6-month intervals, children with chronic OME until the effusion is no longer present, significant hearing loss is identified, or structural abnormalities of the eardrum or middle ear are suspected; (9) should recommend tympanostomy tubes when surgery is performed for OME in a child <4 years old; adenoidectomy should not be performed unless a distinct indication exists (nasal obstruction, chronic adenoiditis); (10) should recommend tympanostomy tubes, adenoidectomy, or both when surgery is performed for OME in a child ≥4 years old; and (11) should document resolution of OME, improved hearing, or improved quality of life when managing a child with OME.

[Russell K](#), [Oliver SE](#), [Lewis L](#), [Barfield WD](#), [Cragan J](#), [Meaney-Delman D](#), [Staples JE](#), [Fischer M](#), [Peacock G](#), [Oduyebo T](#), [Petersen EE](#), [Zaki S](#), [Moore CA](#), [Rasmussen SA](#); **Contributors.**

### **Update: Interim Guidance for the Evaluation and Management of Infants with Possible Congenital Zika Virus Infection - United States, August 2016.**

*MMWR Morb Mortal Wkly Rep.* 2016 Aug 26;65(33):870-878. doi: 10.15585/mmwr.mm6533e2.

**Abstract:** CDC has updated its interim guidance for U.S. health care providers caring for infants born to mothers with possible Zika virus infection during pregnancy (1). Laboratory testing is recommended for 1) infants born to mothers with laboratory evidence of Zika virus infection during pregnancy and 2) infants who have abnormal clinical or neuroimaging findings suggestive of congenital Zika syndrome and a maternal epidemiologic link suggesting possible transmission, regardless of maternal Zika virus test results. Congenital Zika syndrome is a recently recognized pattern of congenital anomalies associated with Zika virus infection during pregnancy that includes microcephaly, intracranial calcifications or other brain anomalies, or eye anomalies, among others (2). Recommended infant laboratory evaluation includes both molecular (real-time reverse transcription-polymerase chain reaction [rRT-PCR]) and serologic (immunoglobulin M [IgM]) testing. Initial samples should be collected directly from the infant in the first 2 days of life, if possible; testing of cord blood is not recommended. A positive infant serum or urine rRT-PCR test result confirms congenital Zika virus infection. Positive Zika virus IgM testing, with a negative rRT-PCR result, indicates probable congenital Zika virus infection. In addition to infant Zika virus testing, initial evaluation of all infants born to mothers with laboratory evidence of Zika virus infection during pregnancy should include a comprehensive physical examination, including a neurologic examination, postnatal head ultrasound, and standard newborn hearing screen. Infants with laboratory evidence of congenital Zika virus infection should have a comprehensive ophthalmologic exam and hearing assessment by auditory brainstem response (ABR) testing before 1 month of age. Recommendations for follow-up of infants with laboratory evidence of congenital Zika virus infection depend on whether abnormalities consistent with congenital Zika syndrome are present. Infants with abnormalities consistent with congenital Zika syndrome should have a coordinated evaluation by multiple specialists within the first month of life; additional evaluations will be needed within the first year of life, including assessments of vision, hearing, feeding, growth, and neurodevelopmental and endocrine function. Families and caregivers will also need ongoing psychosocial support and assistance with coordination of care. Infants with laboratory evidence of congenital Zika virus infection without apparent abnormalities should have ongoing developmental monitoring and screening by the primary care provider; repeat hearing testing is recommended. This guidance will be updated when additional information becomes available.



[Sloan-Heggen CM](#), [Bierer AO](#), [Shearer AE](#), [Kolbe DL](#), [Nishimura CJ](#), [Frees KL](#), [Ephraim SS](#), [Shibata SB](#), [Booth KT](#), [Campbell CA](#), [Ranum PT](#), [Weaver AE](#), [Black-Ziegelbein EA](#), [Wang D](#), [Azaiez H](#), [Smith RJ](#).

### [Comprehensive Genetic Testing in the Clinical Evaluation of 1119 Patients with Hearing Loss.](#)

*Hum Genet.* 2016 Apr;135(4):441-50. doi: 10.1007/s00439-016-1648-8. Epub 2016 Mar 11.

**Abstract:** Hearing loss is the most common sensory deficit in humans, affecting 1 in 500 newborns. Due to its genetic heterogeneity, comprehensive diagnostic testing has not previously been completed in a large multiethnic cohort. To determine the aggregate contribution inheritance makes to non-syndromic hearing loss, we performed comprehensive clinical genetic testing with targeted genomic enrichment and massively parallel sequencing on 1119 sequentially accrued patients. No patient was excluded based on phenotype, inheritance or previous testing. Testing resulted in identification of the underlying genetic cause for hearing loss in 440 patients (39%). Pathogenic variants were found in 49 genes and included missense variants (49%), large copy number changes (18%), small insertions and deletions (18%), nonsense variants (8%), splice-site alterations (6%), and promoter variants (<1%). The diagnostic rate varied considerably based on phenotype and was highest for patients with a positive family history of hearing loss or when the loss was congenital and symmetric. The spectrum of implicated genes showed wide ethnic variability. These findings support the more efficient utilization of medical resources through the development of evidence-based algorithms for the diagnosis of hearing loss.

[Störbeck C](#), [Young A](#).

### [The HI HOPES Data Set of Deaf Children Under the Age of 6 in South Africa: Maternal Suspicion, Age of Identification and Newborn Hearing Screening.](#)

*BMC Pediatr.* 2016 Mar 22;16:45. doi: 10.1186/s12887-016-0574-1.

**Background:** Identification of deafness before 3 months of age substantially improves the socio-linguistic and cognitive development of deaf children. Existing studies demonstrating the feasibility of newborn hearing screening in South Africa have used small samples unrepresentative of general population characteristics. This study establishes the characteristics of the largest data set of deaf infants and their families in South Africa on which there is baseline and longitudinal data (n=532); explores its representativeness in terms of socio-demographic features and reports on access to and quality of newborn hearing screening within the sample. It examines specifically the relationship between age of maternal suspicion of childhood deafness and age of identification of deafness by cohort characteristics.

**Methods:** Secondary analysis, using descriptive and inferential statistics, of a pre-existing longitudinal data set (n=532) of deaf infants under 6 years of age, and their families, collected as routine monitoring of the HI HOPES (HH) early intervention programme.

**Results:** The HH cohort is representative in terms of racial profile and private/public health care use but displays slightly higher level of maternal education and slightly lower socio-economic status than national comparators. 102 out of 532 infants had undergone newborn hearing screening, resulting in 29 true positives, 15 of whom would have met the criteria for targeted screening. Later onset deafness does not account for the 73 false negatives. The median age of maternal suspicion (n=247) of infant deafness was 18 months; the median age of identification of 28 months. Age of identification was unrelated to private/public health care status. The median delay between age of suspicion and age of identification was significantly longer in the public sector (7 m; IQR 0-15 m) compared to the private sector (2 m; IQR 0-8.5 m) (p=0.035). Age of suspicion was unrelated to level of maternal education. Earlier age of suspicion did not predict earlier identification.

**Conclusion:** Targeted screening as timely response to maternal suspicion offers a viable means to reduce substantially the age of identification of deafness in South Africa until implementation of newborn hearing screening on a population-wide basis can be justified.

[Szyfter W](#), [Greczka G](#), [Dąbrowski P](#), [Wróbel M](#).

### [The Report on the Universal Neonatal Hearing Screening Program in Poland between 2003 and 2015.](#)

*Otolaryngol Pol.* 2016 Apr 30;70(2):1-5. doi: 10.5604/00306657.1199346.

**Abstract:** The Universal Neonatal Hearing Screening Program (UNHSP) has been operating in the whole Poland since 2003. Its main goals are to perform a screening hearing in the 2-3 day of life in every newborn baby in Poland and to gather information on risk factors of hearing loss. In total, 505 centers participate in the UNHSP on three reference levels. As of January 19th 2016, the central data base (CDB) of the UNHSP has records of 4,845,036 children, which comprises 96% of all children born in Poland. Hearing loss was diagnosed in 12,974 children, i.e. in 3 out of 1000 children. Here, we present the most important results and conclusions of the UNHSP.

[Uematsu M](#), [Haginoya K](#), [Kikuchi A](#), [Hino-Fukuyo N](#), [Ishii K](#), [Shiihara T](#), [Kato M](#), [Kamei A](#), [Kure S](#).

### [Asymptomatic Congenital Cytomegalovirus Infection with Neurological Sequelae: A Retrospective Study Using Umbilical Cord.](#)

*Brain Dev.* 2016 Oct;38(9):819-26. doi: 10.1016/j.braindev.2016.03.006. Epub 2016 Apr 8.

**Background:** Congenital cytomegalovirus (CMV) infection causes various neurological sequelae. However, most infected infants are asymptomatic at birth, and retrospective diagnosis is difficult beyond the neonatal period.

**Objective:** This study aimed to investigate the aspects of neurological sequelae associated with asymptomatic congenital CMV infection.

**Methods:** We retrospectively analyzed 182 patients who were suspected of having asymptomatic congenital CMV infection with neurological symptoms in Japan. Congenital CMV infection was diagnosed by quantitative polymerase chain reaction amplification of CMV from dried umbilical cord DNA.

**Results:** Fifty-nine patients (32.4%) who tested positive for CMV were confirmed as having congenital CMV infection. Among 54 congenital CMV patients, major neurological symptoms included intellectual disability (n=51, 94.4%), hearing impairment (n=36, 66.7%) and cerebral palsy (n=21, 38.9%), while microcephaly (n=16, 29.6%) and epilepsy (n=14, 25.9%) were less common. In a brain magnetic resonance imaging (MRI) study, cortical dysplasia was observed in 27 CMV-positive patients (50.0%), and all patients (100%) had cerebral white matter (WM) abnormality. Intracranial calcification was detected by CT in 16 (48.5%) of 33 CMV-positive patients. Cerebral palsy, cortical dysplasia and a WM abnormality with a diffuse pattern were associated with marked intellectual disability.

**Conclusion:** Brain MRI investigations are important for making a diagnosis and formulating an intellectual prognosis. Analysis of umbilical cord tissue represents a unique and useful way to retrospectively diagnose congenital CMV infection.

[Van As JW, Van den Berg H, Van Dalen EC.](#)

### [Platinum-Induced Hearing Loss After Treatment for Childhood Cancer.](#)

[Cochrane Database Syst Rev. 2016 Aug 3;\(8\):CD010181. doi: 10.1002/14651858.CD010181.pub2.](#)

**Background:** Platinum-based therapy, including cisplatin, carboplatin, oxaliplatin or a combination of these, is used to treat a variety of paediatric malignancies. Unfortunately, one of the most important adverse effects is the occurrence of hearing loss or ototoxicity. There is a wide variation in the reported prevalence of platinum-induced ototoxicity and the associated risk factors. More insight into the prevalence of and risk factors for platinum-induced hearing loss is essential in order to develop less ototoxic treatment protocols for the future treatment of children with cancer and to develop adequate follow-up protocols for childhood cancer survivors treated with platinum-based therapy.

**Objective:** To evaluate the existing evidence on the association between childhood cancer treatment including platinum analogues and the occurrence of hearing loss.

**Search Methods:** We searched the Cochrane Central Register of Controlled Trials (CENTRAL) (2015, Issue 8), MEDLINE (PubMed) (1945 to 23 September 2015) and EMBASE (Ovid) (1980 to 23 September 2015). In addition, we searched reference lists of relevant articles and the conference proceedings of the International Society for Paediatric Oncology (2008 to 2014), the American Society of Pediatric Hematology/Oncology (2008 to 2015) and the International Conference on Long-Term Complications of Treatment of Children and Adolescents for Cancer (2010 to 2015). Experts in the field provided information on additional studies.

**Selection Criteria:** All study designs, except case reports, case series (i.e. a description of non-consecutive participants) and studies including fewer than 100 participants treated with platinum-based therapy who had an ototoxicity assessment, examining the association between childhood cancer treatment including platinum analogues and the occurrence of hearing loss.

**Data Collection and Analysis:** Two review authors independently performed the study selection. One review author performed data extraction and risk of bias assessment, which was checked by another review author.

**Main Results:** We identified 13 eligible cohort studies including 2837 participants with a hearing test after treatment with a platinum analogue for different types of childhood cancers. All studies had methodological limitations, with regard to both internal (risk of bias) and external validity. Participants were treated with cisplatin, carboplatin or both, in varying doses. The reported prevalence of hearing loss varied considerably between 0% and 90.1%; none of the studies provided data on tinnitus. Three studies reported a prevalence of 0%, but none of these studies provided a definition for hearing loss and there might be substantial or even complete overlap in included participants between these three studies. When only studies that did provide a definition for hearing loss were included, the prevalence of hearing loss still varied widely between 1.7% and 90.1%. All studies were very heterogeneous with regard to, for example, definitions of hearing loss, used diagnostic tests, participant characteristics, (prior) anti-tumour treatment, other ototoxic drugs and length of follow-up. Therefore, pooling of results was not possible. Only two studies included a control group of people who had not received platinum treatment. In one study, the prevalence of hearing loss was 67.1% (95% confidence interval (CI) 59.3% to 74.1%) in platinum-treated participants, while in the control participants it was 7.4% (95% CI 6.2% to 8.8%). However, hearing loss was detected by screening in survivors treated with platinum analogues and by clinical presentation in control participants. It is uncertain what the effect of this difference in follow-up/diagnostic testing was. In the other study, the prevalence of hearing loss was 20.1% (95% CI 17.4% to 23.2%) in platinum-treated participants and 0.4% (95% CI 0.12% to 1.6%) in control participants. As neither study was a randomized controlled trial or controlled clinical trial, the calculation of a risk ratio was not feasible as it is very likely that both groups differed more than only the platinum treatment. Only two studies evaluated possible risk factors using multivariable analysis. One study identified a significantly higher risk of hearing loss in people treated with cisplatin 400 mg/m<sup>2</sup> plus carboplatin 1700 mg/m<sup>2</sup> as compared to treatment with cisplatin 400 mg/m<sup>2</sup> or less, irrespective of the definition of hearing loss. They also identified a significantly higher risk of hearing loss in people treated with non-anthracycline aminoglycosides antibiotics (using a surrogate marker) as compared to people not treated with them, for three out of four definitions of hearing loss. The other study reported that age at treatment (odds ratio less than 1 for each single-unit increase) and single maximum cisplatin dose (odds ratio greater than 1 for each single-unit increase) were significant predictors for hearing loss, while gender was not.

**Authors' Conclusions:** This systematic review shows that children treated with platinum analogues are at risk for developing hearing loss, but the exact prevalence and risk factors remain unclear. There were no data available for tinnitus. Based on the currently available evidence we can only advise that children treated with platinum analogues are screened for ototoxicity in order to make it possible to diagnose hearing loss early and to take appropriate measures. However, we are unable to give recommendations for specific follow-up protocols including frequency of testing. Counselling regarding the prevention of noise pollution can be considered, such as the use of noise-limiting equipment, avoiding careers with excess noise and ototoxic medication. Before definitive conclusions on the prevalence and associated risk factors of platinum-induced ototoxicity can be made, more high-quality research is needed. Accurate and transparent reporting of findings will make it possible for readers to appraise the results of these studies critically.

[Vashistha I, Aseri Y, Singh BK, Verma PC.](#)

### [Prevalence of Hearing Impairment in High Risk Infants.](#)

[Indian J Otolaryngol Head Neck Surg. 2016 Jun;68\(2\):214-7. doi: 10.1007/s12070-015-0869-9. Epub 2015 Jun 30.](#)

**Abstract:** Hearing impairment is prevalent in the general population, early intervention facilitates proper development. The aim of this study was to establish the prevalence of hearing impairment in high-risk infants born between 2013 and 2014. 100 newborns were evaluated using evoked otoacoustic emissions and distortion produce and auditory behavior. Tests were reported if the results were altered. If altered results persisted, the child was referred for impedance testing and when necessary for medical evaluation. Infants referred for BOA and OAE undergone Brainstem auditory evoked potential testing. Of 100 children, 85 children have hearing within normal limits. Hearing impairment was found in 15 out of which 7 had unilateral hearing loss and 8 had bilateral hearing loss. The high prevalence of hearing impairment in this population underlines the importance of early audiological testing.

[Vos B, Senterre C, Lagasse R, Tognola G, Levêque A.](#)

### [Organization of Newborn Hearing Screening Programmes in the European Union: Widely Implemented, Differently Performed.](#)

[Eur J Public Health. 2016 Jun;26\(3\):505-10. doi: 10.1093/eurpub/ckw020. Epub 2016 Mar 23.](#)

**Background:** Implementation of newborn hearing screening programmes is widely recommended and programme organisational designs may differ in practice. The objective of this article was to establish an overview of the newborn hearing screening programmes in the 28 countries of the European Union on four topics (policy-decision, financing, general designs, organisational features).

**Methods:** National or regional programme coordinators completed an online self-administered questionnaire focusing on protocol description and programme organization.

**Results:** Thirty-nine key informants, representing 24 countries, from national or regional levels completed the questionnaire. Newborn hearing screening programmes are or will be shortly implemented largely in the European Union countries. Levels of policy decision-making and organisational decisions are diverse (national, regional or combined). Designs of the programmes (number of steps before diagnosis referral, single

or dual target group protocol) highly varied. However, common organisational elements were observed: hearing screening tests are often performed by nursing staff, in hospitals and early in life. This pattern does not apply when a screening protocol is specifically implemented for newborns with risk factor(s) for hearing impairment or admitted to neonatal intensive care units. Hearing test financing frequently involved public sources, including government and public health funds.

**Conclusion:** Despite the same goal of early identification of hearing-impaired children, there is a high level of diversity in programmes, including policy decisions, financing, general designs and pragmatic organisational choices (e.g. professionals involved, location or time for screening, number of steps in the protocol). Further investigations should analyse these differences in relation to the programmes' contexts and outcomes.

[Voss SE](#), [Herrmann BS](#), [Horton NJ](#), [Amadei EA](#), [Kujawa SG](#).

### [Reflectance Measures from Infant Ears With Normal Hearing and Transient Conductive Hearing Loss.](#)

*Ear Hear.* 2016 Sep-Oct;37(5):560-71. doi: 10.1097/AUD.000000000000293.

**Objective:** The objective is to develop methods to utilize newborn reflectance measures for the identification of middle-ear transient conditions (e.g., middle-ear fluid) during the newborn period and ultimately during the first few months of life. Transient middle-ear conditions are a suspected source of failure to pass a newborn hearing screening. The ability to identify a conductive loss during the screening procedure could enable the referred ear to be either (1) cleared of a middle-ear condition and recommended for more extensive hearing assessment as soon as possible, or (2) suspected of a transient middle-ear condition, and if desired, be rescreened before more extensive hearing assessment.

**Design:** Reflectance measurements are reported from full-term, healthy, newborn babies in which one ear referred and one ear passed an initial auditory brainstem response newborn hearing screening and a subsequent distortion product otoacoustic emission screening on the same day. These same subjects returned for a detailed follow-up evaluation at age 1 month (range 14 to 35 days). In total, measurements were made on 30 subjects who had a unilateral refer near birth (during their first 2 days of life) and bilateral normal hearing at follow-up (about 1 month old). Three specific comparisons were made: (1) Association of ear's state with power reflectance near birth (referred versus passed ear), (2) Changes in power reflectance of normal ears between newborn and 1 month old (maturation effects), and (3) Association of ear's newborn state (referred versus passed) with ear's power reflectance at 1 month. In addition to these measurements, a set of preliminary data selection criteria were developed to ensure that analyzed data were not corrupted by acoustic leaks and other measurement problems.

**Results:** Within 2 days of birth, the power reflectance measured in newborn ears with transient middle-ear conditions (referred newborn hearing screening and passed hearing assessment at age 1 month) was significantly greater than power reflectance on newborn ears that passed the newborn hearing screening across all frequencies (500 to 6000 Hz). Changes in power reflectance in normal ears from newborn to 1 month appear in approximately the 2000 to 5000 Hz range but are not present at other frequencies. The power reflectance at age 1 month does not depend significantly on the ear's state near birth (refer or pass hearing screening) for frequencies above 700 Hz; there might be small differences at lower frequencies.

**Conclusion:** Power reflectance measurements are significantly different for ears that pass newborn hearing screening and ears that refer with middle-ear transient conditions. At age 1 month, about 90% of ears that referred at birth passed an auditory brainstem response hearing evaluation; within these ears the power reflectance at 1 month did not differ between the ear that initially referred at birth and the ear that passed the hearing screening at birth for frequencies above 700 Hz. This study also proposes a preliminary set of criteria for determining when reflectance measures on young babies are corrupted by acoustic leaks, probes against the ear canal, or other measurement problems. Specifically proposed are "data selection criteria" that depend on the power reflectance, impedance magnitude, and impedance angle. Additional data collected in the future are needed to improve and test these proposed criteria.

[Wake M](#), [Ching TY](#), [Wirth K](#), [Poulakis Z](#), [Mensah FK](#), [Gold L](#), [King A](#), [Bryson HE](#), [Reilly S](#), [Rickards F](#).

### [Population Outcomes of Three Approaches to Detection of Congenital Hearing Loss.](#)

*Pediatrics.* 2016 Jan;137(1). doi: 10.1542/peds.2015-1722. Epub 2015 Dec 24.

**Background:** Universal newborn hearing screening was implemented worldwide largely on modeled, not measured, long-term benefits. Comparative quantification of population benefits would justify its high cost.

**Methods:** Natural experiment comparing 3 population approaches to detecting bilateral congenital hearing loss (>25 dB, better ear) in Australian states with similar demographics and services: (1) universal newborn hearing screening, New South Wales 2003-2005, n = 69; (2) Risk factor screening (neonatal intensive care screening + universal risk factor referral), Victoria 2003-2005, n = 65; and (3) largely opportunistic detection, Victoria 1991-1993, n = 86. Children in (1) and (2) were followed at age 5 to 6 years and in (3) at 7 to 8 years. Outcomes were compared between states using adjusted linear regression.

**Results:** Children were diagnosed younger with universal than risk factor screening (adjusted mean difference -8.0 months, 95% confidence interval -12.3 to -3.7). For children without intellectual disability, moving from opportunistic to risk factor to universal screening incrementally improved age of diagnosis (22.5 vs 16.2 vs 8.1 months,  $P < .001$ ), receptive (81.8 vs 83.0 vs 88.9,  $P = .05$ ) and expressive (74.9 vs 80.7 vs 89.3,  $P < .001$ ) language and receptive vocabulary (79.4 vs 83.8 vs 91.5,  $P < .001$ ); these nonetheless remained well short of cognition (mean 103.4, SD 15.2). Behavior and health-related quality of life were unaffected.

**Conclusion:** With new randomized trials unlikely, this may represent the most definitive population-based evidence supporting universal newborn hearing screening. Although outperforming risk factor screening, school entry language still lagged cognitive abilities by nearly a SD. Prompt intervention and efficacy research are needed for children to reach their potential.

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### [Children with Auditory Neuropathy Spectrum Disorder Fitted with Hearing Aids Applying the American Academy of Audiology Pediatric Amplification Guideline: Current Practice and Outcomes.](#)

*J Am Acad Audiol.* 2016 Mar;27(3):204-18. doi: 10.3766/jaaa.15050.

**Background:** Up to 15% of children with permanent hearing loss (HL) have auditory neuropathy spectrum disorder (ANSD), which involves normal outer hair cell function and disordered afferent neural activity in the auditory nerve or brainstem. Given the varying presentations of ANSD in children, there is a need for more evidence-based research on appropriate clinical interventions for this population.

**Purpose:** This study compared the speech production, speech perception, and language outcomes of children with ANSD, who are hard of hearing, to children with similar degrees of mild-to-moderately severe sensorineural hearing loss (SNHL), all of whom were fitted with bilateral hearing aids (HAs) based on the American Academy of Audiology pediatric amplification guidelines.

**Research Design:** Speech perception and communication outcomes data were gathered in a prospective accelerated longitudinal design, with entry into the study between six mo and seven yr of age. Three sites were involved in participant recruitment: Boys Town National Research Hospital, the University of North Carolina at Chapel Hill, and the University of Iowa.

**Study Sample:** The sample consisted of 12 children with ANSD and 22 children with SNHL. The groups were matched based on better-ear pure-tone average, better-ear aided speech intelligibility index, gender, maternal education level, and newborn hearing screening result (i.e., pass or refer).

**Data Collection and Analysis:** Children and their families participated in an initial baseline visit, followed by visits twice a year for children <2 yr of age and once a yr for children >2 yr of age. Paired-sample t-tests were used to compare children with ANSD to children with SNHL.

**Results:** Paired t-tests indicated no significant differences between the ANSD and SNHL groups on language and articulation measures. Children with ANSD displayed functional speech perception skills in quiet. Although the number of participants was too small to conduct statistical analyses for speech perception testing, there appeared to be a trend in which the ANSD group performed more poorly in background noise with HAs, compared to the SNHL group.

**Conclusion:** The American Academy of Audiology Pediatric Amplification Guidelines recommend that children with ANSD receive an HA trial if their behavioral thresholds are sufficiently high enough to impede speech perception at conversational levels. For children with ANSD in the mild-to-severe HL range, the current results support this recommendation, as children with ANSD can achieve functional outcomes similar to peers with SNHL.

[Wu CC](#), [Tsai CH](#), [Hung CC](#), [Lin YH](#), [Lin YH](#), [Huang FL](#), [Tsao PN](#), [Su YN](#), [Lee YL](#), [Hsieh WS](#), [Hsu CJ](#).

### [Newborn Genetic Screening for Hearing Impairment: A Population-Based Longitudinal Study.](#)

*Genet Med.* 2016 Jun 16. doi: 10.1038/gim.2016.66.

**Purpose:** The feasibility of genetic screening for deafness-causing mutations in newborns has been reported in several studies. The aim of this study was to investigate the long-term results in those who screened positive for deafness mutations; these results are crucial to determine the cost-effectiveness to justify population-wide genetic screening.

**Methods:** We performed simultaneous hearing screening and genetic screening targeting four common deafness mutations (p.V37I and c.235delC of GJB2, c.919-2A>G of SLC26A4, and the mitochondrial m.1555A>G) in 5173 newborns at a tertiary hospital between 2009 and 2015. Serial audiometric results up to 6 years old were then analyzed in children with conclusive genotypes.

**Results:** Newborn genetic screening identified 82 (1.6%) babies with conclusive genotypes, comprising 62 (1.2%) with GJB2 p.V37I/p.V37I, 16 (0.3%) with GJB2 p.V37I/c.235delC, and 4 (0.1%) with m.1555A>G. Of these, 46 (56.1%) passed hearing screening at birth. Long-term follow-up demonstrated progressive hearing loss in children with the GJB2 p.V37I/p.V37I and p.V37I/c.235delC genotypes; this hearing loss deteriorated by approximately 1 decibel hearing level (dBHL) per year.

**Conclusion:** We delineated the longitudinal auditory features of the highly prevalent GJB2 p.V37I mutation on a general population basis and confirmed the utility of newborn genetic screening in identifying infants with late-onset or progressive hearing impairment undetectable by newborn hearing screening. *Genet Med* advance online publication 16 June 2016 *Genetics in Medicine* (2016); doi:10.1038/gim.2016.66

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### [A Look into the Crystal Ball for Children Who Are Deaf or Hard of Hearing: Needs, Opportunities, and Challenges.](#)

*Semin Speech Lang.* 2016 Nov;37(4):252-258. Epub 2016 Oct 4.

**Abstract:** Hearing is essential for the development of speech, spoken language, and listening skills. Children previously went undiagnosed with hearing loss until they were 2.5 or 3 years of age. The auditory deprivation during this critical period of development significantly impacted long-term listening and spoken language outcomes. Due to the advent of universal newborn hearing screening, the average age of diagnosis has dropped to the first few months of life, which sets the stage for outcomes that include children with speech, spoken language, and auditory skill testing in the normal range. However, our work is not finished. The future holds even greater possibilities for children with hearing loss.

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### [Follow-Up Results of Newborns after Hearing Screening at a Training and Research Hospital in Turkey.](#)

*J Int Adv Otol.* 2016 Apr;12(1):55-60. doi: 10.5152/iao.2015.1736.

**Objective:** This study aimed to present the follow-up results of newborns after universal newborn hearing screening at a Training and Research Hospital in Istanbul and to determine the ages of diagnosis, hearing aid fitting, and cochlear implantation in newborns with hearing loss.

**Materials and Methods:** A total of 5985 newborns were screened between December 2009 and August 2011 using the transient evoked otoacoustic emission test as the first two steps and automated auditory brainstem response (ABR) test as the third step. Newborns who failed the screening tests were referred to a tertiary hospital for clinic ABR and were followed up at least for 2 years.

**Results:** Of 5985 newborns, 5116 (85.5%) completed the screening. Of 53 newborns who were referred to a tertiary hospital, 13 (0.25%) had a hearing impairment. The mean age of diagnosis, hearing aid fitting, and cochlear implantation were 6.1, 9.5, and 24.5 months, respectively. Among the risk factors for hearing impairment, neonatal intensive care (60%) and consanguineous marriage (50%) were the most common ones that were encountered.

**Conclusion:** Our results were consistent with the national literature. Consanguineous marriage may be a risk factor for hearing impairment where it is commonly practiced because consanguineous marriage is significantly high in parents of deaf children. The ages of diagnosis and hearing aid fitting are still beyond the recommended ages by the Joint Committee on Infant Hearing.