Others’ Publications About EHDI: October 2017 through April 2018

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

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

Of the 72 articles published in other journals from October 2017 through April 2018, most reported on research conducted in countries other than the United States. Clearly, EHDI continues to be a global phenomenon. Staying current with EHDI’s processes and accomplishments requires staying informed about what is happening in other countries. During this time period, the topics most frequently addressed included issues related to the identification or treatment of congenital cytomegalovirus (cCMV) and its relation to childhood hearing loss, the genetics of hearing loss, the use of cochlear implants, language outcomes for children identified in newborn hearing screening programs, and issues related to protocols and procedures used in newborn hearing screening, follow-up, and diagnosis.

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

- Cejas et al. reported that children using cochlear implants perform similarly to hearing peers on measures of intelligence, but those with severe comorbidities are at-risk for cognitive deficits.
- Fitzpatrick et al. in a cross-sectional analysis of 120 children, found that children with unilateral hearing loss tended to perform poorer than those in the mild bilateral hearing loss or normal hearing.
- Grasty et al. followed 348 children who underwent repair of congenital heart disease and found that the prevalence of hearing loss in preschool children after heart surgery in infancy was 20-fold higher than in the 1% prevalence seen in the general population.
- Hao et al. demonstrated that newborn hearing genetic screening could be used in conjunction with newborn hearing screening to identify four genetic mutations frequently associated with congenital or late-onset hearing loss. Genetic mutations were found for 3.01% of the 142,417 neonates screened.
- Rawlinson et al. screened infants for cCMV using PCR of urine and saliva and found ~6% of the large sample of children who had failed a newborn hearing screening test were positive for cCMV. In contrast, Vancor et al., found only 2 of 171 (1.2%) infants who failed newborn hearing screening had confirmed cCMV based on PCR of saliva.
- Zych et al. reported on 14 years of data from the Polish Universal neonatal Hearing Screening Program.

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

Adebanjo T, Godfred-Cato S, Viens L, Fischer M, Staples JE, Kuhnert-Tallman W, Walke H, Oduyebo T, Polen K, Peacock G, Meaney-Delman D, Honein MA, Rasmussen SA, Moore CA Update: Interim Guidance for the Diagnosis, Evaluation, and Management of Infants with Possible Congenital Zika Virus Infection - United States, October 2017. MMWR Morb Mortal Wkly Rep. 2017 Oct 20;66(41):1089-1099. doi: 10.15585/mmwr.mm6641a1. CDC has updated its interim guidance for U.S. health care providers caring for infants with possible congenital Zika virus infection (1) in response to recently published updated guidance for health care providers caring for pregnant women with possible Zika virus exposure (2), unknown sensitivity and specificity of currently available diagnostic tests for congenital Zika virus infection, and recognition of additional clinical findings associated with congenital Zika virus infection. All infants born to mothers with possible Zika virus exposure* during pregnancy should receive a standard evaluation at birth and at each subsequent well-child visit including a comprehensive physical examination, age-appropriate vision screening and developmental monitoring and screening using validated tools (3-5), and newborn hearing screen at birth, preferably using auditory brainstem response (ABR) methodology (6). Specific guidance for laboratory testing and clinical evaluation are provided for three clinical scenarios in the setting of possible maternal Zika virus exposure: 1) infants with clinical findings consistent with congenital Zika syndrome regardless of maternal testing results, 2) infants without clinical
findings consistent with congenital Zika syndrome who were born to mothers with laboratory evidence of possible Zika virus infection,† and 3) infants without clinical findings consistent with congenital Zika syndrome who were born to mothers without laboratory evidence of possible Zika virus infection. Infants in the first two scenarios should receive further testing and evaluation for Zika virus, whereas for the third group, further testing and clinical evaluation for Zika virus are not recommended. Health care providers should remain alert for abnormal findings (e.g., postnatal-onset microcephaly and eye abnormalities without microcephaly) in infants with possible congenital Zika virus exposure without apparent abnormalities at birth.

Appelbaum EN, Howell JB, Chapman D, Pandya A, Dodson KM. Analysis of risk factors associated with unilateral hearing loss in children who initially passed newborn hearing screening. Int J Pediatr Otorhinolaryngol. 2018 Mar;106:100-104. doi: 10.1016/j.ijporl.2018.01.024. Epub 2018 Feb 2. OBJECTIVE: To analyze 2007 Joint Committee on Infant Hearing (JCIH) risk factors in children with confirmed unilateral hearing loss (UHL) who initially passed newborn hearing screening. METHODS: Retrospective record review of 16,108 infants who passed newborn hearing screening but had one or more JCIH risk factors prompting subsequent follow-up through the universal newborn hearing screening (UNHS) program in Virginia from 2010 to 2012. The study was reviewed and qualified as exempt by the Virginia Commonwealth University Institutional Review Board (IRB) and the Virginia Department of Health. RESULTS: Over the 2-year study period, 14896 (4.9% of total births) children passed UNHS but had the presence of one or more JCIH risk factor. Ultimately, we identified 121 babies from this group with confirmed hearing loss (0.7%), with 48 babies (0.2%) showing UHL. The most common risk factors associated with the development of confirmed UHL after passing the initial screen were neonatal indicators, craniofacial anomalies, family history, and stigmata of syndrome associated with hearing loss. CONCLUSION: Neonatal indicators and craniofacial anomalies were the categories most often found in children with confirmed unilateral hearing loss who initially passed their newborn hearing screen. While neonatal indicators were also the most common associated risk factor in all hearing loss, craniofacial abnormalities are relatively more common in children with UHL who initially passed newborn hearing screening. Further studies assessing the etiology underlying the hearing loss and risk factor associations are warranted.

Bianchin G, Tribi L, Formigoni P, Russo C, Polizzi V. Sequential pediatric bilateral cochlear implantation: The effect of time interval between implants. Int J Pediatr Otorhinolaryngol. 2017 Nov;102:10-14. doi: 10.1016/j.ijporl.2017.08.025. Epub 2017 Aug 25. OBJECTIVE: To examine speech intelligibility in children subjected to sequential bilateral cochlear implants (CI) surgery and to assess the influence of the inter-stage interval duration. INTRODUCTION: Binaural hearing recovery can have additional benefits, especially in speech and language development in patients with congenital profound sensorineural hearing loss; so recently there has been an increase in the number of children receiving bilateral CI. METHODS: Twenty-seven children who underwent sequential bilateral cochlear implant (SBCI) with a short (1-3 yrs), medium (4-6 yrs) and long (7-12 yrs) range interval between both implantations, respectively, were evaluated. All patients underwent periodic speech perception test in quiet and noise after second implant activation in three conditions: with the first or second implant alone and with both implants. Results were examined according to the inter-stage interval. RESULTS: Speech intelligibility in noise was significantly better under bilateral conditions than either ear alone, in all three groups. Small improvements were seen in quiet, especially in the third group (6-12 yrs). CONCLUSION: Benefits of second implant in the early-implanted children and after a short inter-implant delay are more evident. However our study support that, even after a long period of deafness and despite a prolonged inter-stage interval, sequential bilateral cochlear implantation should be considered. LEVEL OF EVIDENCE: Level 4. DISCUSSION: Prophylactic HIG administration in pregnant women after CMV primary infection seems not to reduce significantly the rate of congenital infection, but is safe and it could have a favorable effect on the symptoms and sequelae of infected fetuses. The risk of long-term sequelae in fetuses without US abnormalities before HIG is low, so it could be an option in infected fetuses with normal imaging. On the other hand, the risk of sequelae among infected fetuses with abnormalities in fetal ultrasonography before HIG despite treatment is high.

Bostic K, Lewis RM, Chai B, Manganella JL, Barrett DL, Kawai K, Kenna MA, Stiles DJ, Clark T. Enlarged Vestibular Aqueduct and Cochlear Implants: The Effect of Early Counseling on the Length of Time Between Candidacy and Implantation. Otol Neurotol. 2018 Feb;39(2):e90-e95. doi: 10.1097/MAO.0000000000001663. OBJECTIVE: To determine if discussing cochlear implantation (CI) with patients with enlarged vestibular aqueducts (EVA) and their families before reaching audiological criteria for CI candidacy affects the length of time between reaching audiological candidacy and CI surgery, and to describe the universal newborn hearing screening (UNHS) results and communication modality in this sample. PATIENTS: Forty-two patients (25 females) with confirmed EVA and cochlear implants.
INTERVENTION(S): Diagnostic CI visit.
MAIN OUTCOME MEASURES: The primary outcome measure is the difference in length of time between reaching audiological candidacy for CI and surgical implantation between those who had preliminary discussions regarding CI with their medical and healthcare providers before reaching audiological candidacy versus who had discussions after reaching candidacy. The secondary outcome measure is the result of the UNHS and primary mode of communication used by each patient.
RESULTS: Discussing CI before reaching audiological candidacy was associated with a significantly shorter duration between reaching audiological candidacy and receiving CI (median=3.1 mo; interquartile range [IQR]=1.7-5.4) as compared with discussing CI after reaching candidacy (median=5.8 mo; IQR=3.2-11.2; p=0.012). Participants born after the implementation of the UNHS, 16 of 24 patients referred on one or both ears. Communication modalities were evenly divided between utilizing sign-support English and oral/aural communicators only.
CONCLUSIONS: Discussion of CI in patients with EVA before reaching audiological candidacy reduces the amount of time the child is without adequate auditory access and contributes to a constructive and interactive preparatory experience.

BACKGROUND: Early detection and appropriate intervention for children with hearing impairment is important for maximizing functioning and quality of life. The lack of ear and hearing services in low income countries is a significant challenge, however, evidence suggests that even where such services are available, and children are referred to them, uptake is low. The aim of this study was to assess uptake of and barriers to referrals to ear and hearing services for children in Thyolo District, Malawi.
METHODS: This was a mixed methods study. A survey was conducted with 170 caregivers of children who were referred for ear and hearing services during community-based screening camps to assess whether they had attended their referral and reasons for non-attendance. Semi-structured interviews were conducted with 23 caregivers of children who did not take up their referral to explore in-depth the reasons for non-uptake. In addition, 15 stakeholders were interviewed. Thematic analysis of the interview data was conducted and emerging trends were analysed.
RESULTS: Referral uptake was very low with only 5 out of 150 (3%) children attending. Seven main interacting themes for non-uptake of referral were identified in the semi-structured interviews: location of the hospital, lack of transport, other indirect costs of seeking care, fear and uncertainty about the referral hospital, procedural problems within the camps, awareness and understanding of hearing loss, and lack of visibility and availability of services.
CONCLUSION: This study has highlighted a range of interacting challenges faced by families in accessing ear and hearing services in this setting. Understanding these context specific barriers to non-uptake of ear and hearing services is important for designing appropriate interventions to increase uptake.

Cejas I, Mitchell CM, Hoffman M, Quittner AL; and the CDaCI Investigative Team. Comparisons of IQ in Children With and Without Cochlear Implants: Longitudinal Findings and Associations With Language. Ear Hear. 2018 Apr 5. doi: 10.1097/AUD.0000000000000578. [Epub ahead of print]
OBJECTIVES: To make longitudinal comparisons of intelligence quotient (IQ) in children with cochlear implants (CIs) and typical hearing peers from early in development to the school-age period. Children with additional comorbidities and CIs were also evaluated. To estimate the impact of socioeconomic status and oral language on school-age cognitive performance.
DESIGN: This longitudinal study evaluated nonverbal IQ in a multicenter, national sample of 147 children with CIs and 75 typically hearing peers. IQ was evaluated at baseline, prior to cochlear implantation, using the Bayley Scales of Infant and Toddler Development and the Leiter International Performance Scale. School-age IQ was assessed using the Wechsler Intelligence Scales for Children. For the current study, only the Perceptual Reasoning and Processing Speed indices were administered. Oral language was evaluated using the Comprehensive Assessment of Spoken Language.
RESULTS: Children in the CI group scored within the normal range of intelligence at both time points. However, children with additional comorbidities scored significantly worse on the Processing Speed, but not the Perceptual Reasoning Index. Maternal education and language were significantly related to school-age IQ in both groups. Importantly, language was the strongest predictor of intellectual functioning in both children with CIs and normal hearing.
CONCLUSION: These results suggest that children using cochlear implants perform similarly to hearing peers on measures of intelligence, but those with severe comorbidities are at-risk for cognitive deficits. Despite the strong link between socioeconomic status and intelligence, this association was no longer significant once spoken language performance was accounted for. These results reveal the important contributions that early intervention programs, which emphasize language and parent training, contribute to cognitive functioning in school-age children with CIs. For families from economically disadvantaged backgrounds, who are at-risk for suboptimal outcomes, these early intervention programs are critical to improve overall functioning.

Objective: To investigate the effects in adolescence of bilateral permanent childhood hearing loss (PCHL) > 40 dB and of exposure to universal newborn hearing screening (UNHS) on societal costs accrued over the preceding 12 months.

Design setting participants: An observational cohort study of a sample of 110 adolescents aged 13-20 years, 73 with PCHL and 37 in a normally hearing comparison group (HCG) closely similar in respect of place and date of birth to those with PCHL, drawn from a 1992-1997 cohort of 157 000 births in Southern England, half of whom had been exposed to a UNHS programme.

Intervention: Birth in periods with and without UNHS. Outcome measures: Resource use and costs in the preceding 12-month period, estimated from interview at a mean age of 16.9 years and review of medical records. Effects on costs were examined in regression models.

Results: Mean total costs for participants with PCHL and the HCG were £15 914 and £5 883, respectively (difference £10 031, 95% CI £6 460 to £13 603), primarily driven by a difference in educational costs. Compared with the HCG, additional mean costs associated with PCHL of moderate, severe and profound severity were £5 916, £6 605 and £18 437, respectively. The presence of PCHL and an additional medical condition (AMC) increased costs by £15 385 (95% CI £8 532 to £22 238). An increase of one unit in receptive language z-score was associated with £1 161 (95% CI £8 424 to £23 89) lower costs. Birth during periods of UNHS was not associated with significantly lower overall costs (difference £3594, 95% CI -£2981 to £10 106).

Conclusions: The societal cost of PCHL was greater with more severe losses and in the presence of AMC and was lower in children with superior language scores. There was no statistically significant reduction in costs associated with birth in periods with UNHS. Trial registration number: ISRCTN03307358, pre-results.


Purpose: Early auditory experiences are fundamental in infant language acquisition. Research consistently demonstrates the benefits of early intervention (i.e., hearing aids) to language outcomes in children who are deaf and hard of hearing. The nature of these benefits and their relation with prefitting development are, however, not well understood.

Method: This study examined Ontario Infant Hearing Program birth cohorts to explore predictors of performance on the Preschool Language Scale-Fourth Edition at the time of (N = 47) and after (N = 19) initial hearing aid intervention.

Results: Regression analyses revealed that, before the hearing aid fitting, severity of hearing loss negatively predicted 19% and 10% of the variance in auditory comprehension and expressive communication, respectively. After hearing aid fitting, children’s standard scores on language measures remained stable, but they made significant improvement in their progress values, which represent individual skills acquired on the test, rather than standing relative to same-age peers. Magnitude of change in progress values was predicted by a negative interaction of prefitting language ability and severity of hearing loss for the Auditory Comprehension scale.

Conclusions: These findings highlight the importance of considering a child’s prefitting language ability in interpreting eventual language outcomes. Possible mechanisms of hearing aid benefit are discussed.

Supplemental Materials: https://doi.org/10.23641/asha.5538868.


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

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

RESULTS: Overall pass rate was 87.5% (n = 264; 231 passes vs. 33 fails). Of these 84.0% of hospital recruited children passed (n = 156; 131 passes vs. 25 fails), compared to 92.6% of community recruitments (n = 108; 100 passes vs. 8 fails), which was significantly different p = 0.039). If analyzed by age groups, this difference was only observed in children younger than 6 years (p = 0.007).
CONCLUSION: Hospitals as recruitment sites for healthy controls seem to affect TEOAE pass rates. We advise for a cautious approach when recruiting healthy TEOAE control collectives under the age of 6 in a hospital setting. In children older than 6 years conventional pure-tone audiometry remains the standard method for hearing screening.


INTRODUCTION: In children with bilateral severe to profound hearing loss, bilateral hearing can be achieved by either bimodal stimulation (CIHA) or bilateral cochlear implantation (BICI). The aim of this study was to analyse the audiolologic test protocol that is currently applied to make decisions regarding the bilateral hearing modality in the paediatric population.

METHODS: Pre- and postoperative audiolologic test results of 21 CIHA, 19 sequential BICI and 12 simultaneous BICI children were examined retrospectively.

RESULTS: Deciding between either simultaneous BICI or unilateral implantation was mainly based on the infant’s preoperative Auditory Brainstem Response thresholds. Evolution from CIHA to sequential BICI was mainly based on the audiometric test results in the contralateral (hearing aid) ear after unilateral cochlear implantation. Preoperative audiometric thresholds in the hearing aid ear were significantly better in CIHA versus sequential BICI children (p < 0.001 and p = 0.001 in unaided and aided condition, respectively). Decisive values obtained in the hearing aid ear in favour of BICI were: An average hearing threshold measured at 0.5, 1, 2 and 4 kHz of at least 93 dB HL without, and at least 52 dB HL with hearing aid together with a 40% aided speech recognition score and a 70% aided score on the phoneme discrimination subtest of the Auditory Speech Sounds Evaluation test battery.

CONCLUSIONS: Although pure tone audiometry offers no information about bimodal benefit, it remains the most obvious audiomteric evaluation in the decision process on the mode of bilateral stimulation in the paediatric population. A theoretical test protocol for adequate evaluation of bimodal benefit in the paediatric population is proposed.


OBJECTIVES: Children with unilateral hearing loss (UHL) are being diagnosed at younger ages because of newborn hearing screening. Historically, they have been considered at risk for difficulties in listening and language development. Little information is available on contemporary cohorts of children identified in the early months of life. We examined auditory and language acquisition outcomes in a contemporary cohort of early-identified children with UHL and compared their outcomes at preschool age with peers with mild bilateral loss and with normal hearing.

DESIGN: As part of the Mild and Unilateral Hearing Loss in Children Study, we collected auditory and spoken language outcomes on children with unilateral, bilateral hearing loss and with normal hearing over a four-year period. This report provides a cross-sectional analysis of results at age 48 months. A total of 120 children (38 unilateral and 31 bilateral mild, 51 normal hearing) were enrolled in the study from 2010 to 2015. Children started the study at varying ages between 12 and 36 months of age and were followed until age 36-48 months. The median age of identification of hearing loss was 3.4 months (IQR: 2.0, 5.5) for unilateral and 3.6 months (IQR: 2.7, 5.9) for the mild bilateral group. Families completed an intake form at enrolment to provide baseline child and family-related characteristics. Data on amplification fitting and use were collected via parent questionnaires at each annual assessment interval. This study involved a range of auditory development and language measures. For this report, we focus on the end of follow-up results from two auditory development questionnaires and three standardized speech-language assessments. Assessments included in this report were completed at a median age of 47.8 months (IQR: 38.8, 48.5). Using ANOVA, we examined auditory and language outcomes in children with UHL and compared their scores to children with mild bilateral hearing loss and those with normal hearing.

RESULTS: On most measures, children with UHL performed poorer than those in the mild bilateral and normal hearing study groups. All children with hearing loss performed at lower levels compared to the normal hearing control group. However, mean standard scores for the normal hearing group in this study were above normative means for the language measures. In particular, children with UHL showed gaps compared to the normal hearing control group in functional auditory listening and in receptive and expressive language skills (three quarters of one standard deviation below) at age 48 months. Their performance in receptive vocabulary and speech production was not significantly different from that of their hearing peers.

CONCLUSIONS: Even when identified in the first months of life, children with UHL show a tendency to lag behind their normal hearing peers in functional auditory listening and in receptive and expressive language development.
OBJECTIVES: To compare the results after cochlear implantation achieved by monolingual and bilingual deaf children implanted at our Institution, with the aim of understanding if there are any differences between the two groups and if there is a correlation between the outcomes and some patients’ variables.

METHODS: The study group was composed by 14 bilingual deaf children and the control group by the same number of monolingual children implanted at our Institution. The control group was obtained by matching to each bilingual child a monolingual one with a similar clinical history regarding age at hearing loss diagnosis, age at first hearing-aids fitting and age at CI procedure. Children received a speech perception and linguistic development evaluation through specific structured tests. The linguistic competence of the patients both in mainstream and native language was determined by the Student Oral Language Observation Matrix (SOLOM).

RESULTS: We did not find any statistically significant differences between bilingual and monolingual children in speech perception outcomes. Nevertheless, we obtained different results concerning language skills: bilingual implanted children scored lower at structured language tests, even if the difference was not statistically relevant. Bilingual children scored significantly lower than monolingual ones at the SOLOM scale for linguistic competence.

CONCLUSION: The results reported in the present study show better language skills after cochlear implant in Italian monolingual cases than in bilingual ones. This seems to be related to the condition of bilingualism in Italy, mainly related to immigration, and frequently associated with low socio-economic levels, poor competence in the mainstream language and poor social integration, with a suboptimal exposure to the mainstream language and difficulties in following the rehabilitative program.

How does a bilingual environment affect the results in children with cochlear implants compared to monolingual-matched children? An Italian follow-up study.

Fowler KB, Boppana SB.
Congenital cytomegalovirus infection.

Funamura JL.
Evaluation and management of nonsyndromic congenital hearing loss.

Ghadersohi S, Ida JB, Bhushan B, Billings KR.
Outcomes of tympanoplasty in children with down syndrome.

RECENT FINDINGS: The prevalence of chronic otitis media with effusion (COME), and Eustachian tube dysfunction (ETD) is high in Down syndrome (DS) patients. This often necessitates multiple tympanostomy tube (TT) placements resulting in a higher rate of persistent tympanic membrane (TM) perforation requiring tympanoplasty for repair.

OBJECTIVES: To assess risk factors for persistent perforation and outcomes of tympanoplasty in DS patients.

METHODS: Retrospective case series of 91 ears in 69 DS patients with TM perforations, who were either observed or underwent tympanoplasty. Clinical features, surgical outcomes, and hearing outcomes were assessed.

RESULTS: 91 ears were evaluated. Sixty perforations were observed, and 31 perforations were repaired. The closure rate was 54.8% for primary surgery, and 70.9% after secondary surgical interventions in the Tympanoplasty Group, compared to 33.0% spontaneous closure rate in the Observation Group (p < 0.001). The only risk factor for failed tympanoplasty repair was persistent COME/ETD (OR 27.2, p = 0.001). In the Observation Group perforations diagnosed at an
older age, with >3 TT insertions, and with persistent COME/ETD were less likely to close spontaneously. Patients undergoing tympanoplasty had worse preoperative pure tone averages than those being observed, but significant improvement in air-bone gaps were noted in the Tympanoplasty Group (p = 0.02) post-operatively. Patients were often rehabilitated with hearing aids regardless of intervention (53.3% Observation Group, 48.4% Tympanoplasty Group).

CONCLUSIONS: Persistent TM perforation in children with Down syndrome was associated with a history of COME/ETD, and multiple prior TT insertions. Tympanoplasty was successful for repair in most patients who underwent surgical intervention, but residual hearing loss was common.


BACKGROUND: Natural history and long term prognosis of congenital cytomegalovirus (CMV) disease according to maternal primary versus non-primary infection are not clearly documented.

OBJECTIVE: To investigate clinical, laboratory and neuroimaging features at onset and long term outcome of congenitally CMV-infected patients born to mothers with non-primary infection compared with a group of patients born to mothers with primary infection.

STUDY DESIGN: Consecutive neonates born from 2002 to 2015 were considered eligible for the study. Patients underwent clinical, laboratory and instrumental investigation, and audiologic and neurodevelopmental evaluation at diagnosis and during the follow up.

RESULTS: A cohort of 158 congenitally infected children was analyzed. Ninety-three were born to mothers with primary CMV infection (Group 1) and 65 to mothers with a non-primary infection (Group 2). Eighty-eight infants had a symptomatic congenital CMV disease: 49 (46.2%) in Group 1 and 39 (60%) in Group 2. Maternal and demographic characteristics of patients of Group 1 and Group 2 were comparable, with the exception of prematurity and a 1-min Apgar score less than 7, which were more frequent in Group 2 compared to Group 1. Prevalence of neuroimaging findings did not significantly differ between the two groups. An impaired neurodevelopmental outcome was observed in 23.7% of patients of Group 1 and in 24.6% cases of Group 2. Similarly, the frequency of hearing loss did not differ between the two groups (25.8% versus 26.2%, respectively).

CONCLUSIONS: Neurodevelopmental and hearing sequelae are not affected by the type of maternal CMV infection. Preventing strategies should be developed for both primary and non-primary infections.

Goh BS, Fadzilah N, Abdullah A, Othman BF, Umat C.


OBJECTIVES: Cochlear implant (CI) greatly enhances auditory performance as compared to hearing aids and has dramatically affected the educational and communication outcomes for profoundly deaf children. Universiti Kebangsaan Malaysia (UKM) pioneered CI program in 1995 in the South East Asia. We would like to report the long-term outcomes of UKM paediatric cochlear implantation in terms of: the proportion of children who were implanted and still using the device, the children’s modes of communication, their educational placements, and their functional auditory/oral performance. We also examined the factors that affected the outcomes measured.

STUDY DESIGN: This was a cross sectional observational study. METHODS: Two sets of questionnaires were given to 126 parents or primary caregivers of the implantees. The first set of questionnaire contained questions to assess the children’s usage of CI, their types of education placement, and their modes of communication. The second set of questionnaire was the Parent’s Evaluation Of Aural/Oral Performance of Children (PEACH) to evaluate the children’s auditory functionality.

RESULTS: Our study showed that among the implantees, 97.6% are still using their CI, 69.8% communicating orally, and 58.5% attending mainstream education. For implantees that use oral communication and attend mainstream education, their mean age of implantation is 38 months. This is significantly lower compared to the mean age of implantation of implantees that use non-oral communication and attend non-mainstream education. Simple logistic regression analysis shows age of implantation reliably predicts implantees (N = 126) would communicate using oral communication with odds ratio of 0.974, and also predict mainstream education (N = 118) with odds ratio of 0.967. The median score of PEACH rating scale is 87.5% in quiet, and this significantly correlates with an earlier age of implantation (r = -0.235 p = 0.048).

CONCLUSIONS: UKM Cochlear Implant Program has achieved reasonable success among the pediatric implantees, with better outcomes seen in those implanted at the age of less than 4 years old.
Grandpierre V, Fitzpatrick EM, Na E, Mendonca O.
School-aged Children with Mild Bilateral and Unilateral Hearing Loss: Parents’ Reflections on Services, Experiences, and
Outcomes.
Following the establishment of newborn hearing screening programs, age of identification and length of time before
receiving interventions has been reduced for children, including those with milder degrees of hearing loss who were
previously not identified until school age. This population of early-identified children requires new support programs for
parents. Although literature is emerging on how parents experience the initial years, there is limited information on sup-
port needs during early school years. The objectives were to gain insights into parents’ experiences with services during
the early period of identification until early school years, as well as their perceptions of the consequences of hearing loss
on their child's overall development. A qualitative research design informed by Interpretive Description was employed.
Individual semi-structured interviews were conducted with 12 parents of children identified with mild hearing loss. Trans-
cripts were analyzed using a constant comparative method. Four themes emerged from the data: early experiences with services and hearing technology, effects of hearing loss on social functioning, effects of hearing loss on language and academics, and experiences in early school years. From parents’ perspectives, more support during the early school years is needed to help ensure academic success.

Hearing Loss after Cardiac Surgery in Infancy: An Unintended Consequence of Life-Saving Care.
OBJECTIVES: To investigate the prevalence of hearing loss after cardiac surgery in infancy, patient and operative factors associated with hearing loss, and the relationship of hearing loss to neurodevelopmental outcomes.
STUDY DESIGN: Audiologic and neurodevelopmental evaluations were conducted on 348 children who underwent repair of congenital heart disease at the Children’s Hospital of Philadelphia as part of a prospective study evaluating neurodevelopmental outcomes at 4 years of age. A prevalence estimate was calculated based on presence and type of hearing loss. Potential risk factors and the impact of hearing loss on neurodevelopmental outcomes were evaluated.
RESULTS: The prevalence of hearing loss was 21.6% (95% CI, 17.2-25.9). The prevalence of conductive hearing loss, sensorineural hearing loss, and indeterminate hearing loss were 12.4% (95% CI, 8.8-16.0), 6.9% (95% CI, 4.1-9.7), and 2.3% (95% CI, 0.6-4.0), respectively. Only 18 of 348 subjects (5.2%) had screened positive for hearing loss before this study and 10 used a hearing aid. After adjusting for patient and operative covariates, younger gestational age, longer postoperative duration of stay, and a confirmed genetic anomaly were associated with hearing loss (all P < .01). The presence of hearing loss was associated with worse language, cognition and attention (P < .01).
CONCLUSIONS: These findings suggest that the prevalence of hearing loss in preschool children after heart surgery in infancy may be 20-fold higher than in the 1% prevalence seen in the general population. Younger gestational age, presence of a genetic anomaly, and longer postoperative duration of stay were associated with hearing loss. Hearing loss was associated with worse neurodevelopmental outcomes.

Greczka G, Zych M, Wróbel M, Dąbrowski P, Szyfter Harris J, Szyfter W.
Analysis of follow-up at the diagnostic level in the Polish Universal Neonatal Hearing Screening Programme.
Objectives Routine analysis showed that between 1 June and 30 November 2014, only 47.6% of expected follow-up visits at the diagnostic level were registered in the Polish Universal Neonatal Hearing Screening Programme central database. We attempted to detect and analyse the reasons for this low percentage.
Methods A telephone survey questionnaire was developed for parents whose children had not registered for consultation at the diagnostic level, or had not received a final diagnosis according to the programme database. Questions aimed to verify the database records and compare these with information received from and given to parents. From the 7888 children not registered at the diagnostic level, 3239 records were randomly selected, i.e. 52.4% of those who had been expected to attend.
Results Questions were answered by 1950 parents (60.2% of the selected group). Of these, 52.1% (n = 734) had attended for diagnostic tests, but this was not recorded in the database. The most common reasons for not attending were the long waiting time for the visit (36.09%), lack of referral to a visit (25.9%) and conscious parent decision (16.35%).
Conclusion The telephone survey disclosed omissions in database registration, and that in fact 83.6% of children had attended at the diagnostic level.
Large scale newborn deafness genetic screening of 142,417 neonates in Wuhan, China.
Almost one third of the three million people in China suffering severe deafness are children, and 50% of these cases are believed to have genetic components to their etiology. Newborn hearing genetic screening can complement Universal Neonatal Hearing Screening for the diagnosis of congenital hearing loss as well as identifying children at risk for late-on-
set and progressive hearing impairment. The aim of this joint academic and Ministry of Health project was to prototype a cost effective newborn genetic screen in a community health setting on a city-wide level, and to ascertain the prevalence of variation at loci that have been associated with non-syndromic hearing loss. With the participation of 143 local hospitals in the city of Wuhan, China we screened 142,417 neonates born between May 2014 and Dec. 2015. The variants GJB2 c.235delC, SLC26A4 c.919-2A>G, and mitochondrial variants m.1555A>G and m.1494C>T were assayed using real time PCR. Newborns found to carry a variant were re-assayed by sequencing in duplicate. Within a subset of 707 newborns we assayed using real-time PCR and ARMS-PCR to compare cost, sensitivity and operating procedure. The most frequent hearing loss associated allele detected in this population was the 235delC variant in GJB2 gene. In total, 4289 (3.01%) newborns were found to carry at least one allele of either GJB2 c.235delC, SLC26A4 c.919-2A>G or two assayed MT-RNR1 variants. There was complete accordance between the real-time PCR and the ARMS PCR, though the real-time PCR had a much lower failure rate. Real-time PCR had a lower cost and operating time than ARMS PCR. Ongoing collaboration with the participating hospitals will determine the specificity and sensitivity of the association of the variants with hearing loss at birth and arising in early childhood, allowing an estimation of the benefits of newborn hearing genetic screening in a large-scale community setting.


Evaluation of middle ear function in infants is the key to distinguish sensorineural hearing loss and conductive hearing loss, and acoustic immittance test is the routine audiological evaluation of middle ear function. Because of the characteristics of middle ear in infants, middle ear examination parameters of adults are not suitable for infants. This article reviewed the current multiple acoustic immittance methods for detecting middle ear function in infants, and summarized the advantages and disadvantages of these methods.


**Purpose:** This study screens for deafness gene mutations in newborns in the Northwest China population.

**Method:** The 9 sites of 4 common deafness genes (GJB2, GJB3, SLC26A4, and mt 12S rRNA) were detected by bloodspot-based gene chip array in 2,500 newborns.

**Results:** We detected mutations of the 4 genes in 101 (4.04%) newborns; particularly, 0.20% detected the double mutations. In the Hui population, 4.58% of the newborns tested positive for mutations, whereas 4.01% of Han newborns tested positive for mutations. The detection rates are as follows: 1.44% for GJB2 235delC, 1.08% for SLC26A4 IVS7-2A>G, 0.48% for GJB2 299_300delAT, 0.28% for SLC26A4 2168A>G, 0.2% for mt 12S rRNA 1555A>G, and 0.16% for GJB3 538C>T. The 31.25% (5/16) of infants with GJB2 235delC, 50% (3/6) with GJB2 299_300delAT, and 25% (3/12) with SLC26A4 IVS7-2A>G showed abnormal hearing when tested; only 1 double mutation case received the hearing test, and this infant showed abnormality in both ears on the hearing test.

**Conclusions:** High mutation rates in the common deafness genes were detected in newborns in Northwest China. Our study is helpful in understanding the deafness genomic epidemiology and also provides evidence for prenatal and postnatal care as well as policy making on population health in the region.


**Aim** To identify risk factors for hearing impairment presented in neonates born in Cantonal Hospital Zenica (CHZ) and to estimate their influence on outcome of hearing tests in Newborn Hearing Screening (NHS).

**Methods** Retrospective-prospective study was done at the Department of Gynaecology and Maternity. The NHS was performed with transitory evoked otoacoustic emissions (TEOAE) during a six-month period using “Titan” device (Interacoustics, Denmark). The questionnaire was written for the purpose of getting more structured basic information about every newborn and to identify risk factors for hearing impairment. Chi-square test was used to investigate the difference between experimental and control group refer incidence.

**Results** A total of 1217 newborns was screened for hearing impairment of which 259 (21.28%) with one or more known risk factors for hearing impairment. The following risk factors for hearing impairment were identified during the study period: family history of permanent childhood hearing impairment in 42 (3.45%) newborns, prematurity in 39 (3.21%), low APGAR scores in 29 (2.40%), asphyxia in 31 (2.55%), hyperbilirubinemia in 41 (3.37%), admission of ototoxic medication (aminoglycosides) after birth in 155 (12.74%).

**Conclusion** There were many serious risk factors for hearing loss identified in this study. Identification of risk factors for hearing impairment in neonates is necessary because a follow up of the children with risk factors is very important.
Huang MP, Sheng HB, Ren Y, Li Y, Huang ZW, Wu H.

[Effects of bimodal intervention on the development of auditory and speech ability in infants with unilateral cochlear implantation].


Objective: To explore the effects of bimodal intervention on the development of auditory and speech ability in the infants with unilateral cochlear implantation(CI).

Methods: Total 35 bilateral profound sensorineural hearing loss infants with unilateral CI, aged 0.7 to 2.8 years old, were selected. The subjects were divided into two groups: the group with unilateral CI(cochlear implant alone, n= 15), and the bimodal group with CI and contralateral fitting hearing aid(n= 20). Their auditory and speech abilities were estimated at the different time points after switch-on(the 0th, 0.5th, 1st, 3rd, 6th, 12th, 18th, and 24th month, respectively) using Infant Toddler-Meaningful Auditory Integration Scale(IT-MAIS), Meaningful Use of Speech Scale(MUSS), Categories of Auditory Performance(CAP), and Speech Intelligibility Rating(SIR) scores.

Results: The IT-MAIS scores of bimodal group after switch-on were higher than unilateral CI group(the 0.5th, 1st, 3rd, 6th, 12th, 18th and 18th month), the statistical significances were identified at the 0.5th, 1st, 3rd, 6th, and 12th month, respectively(P<0.05). The CAP scores of bimodal group before CI operation and after switch-on(the 0.5th, 1st, 3rd, 6th, 12th, 18th and 24th month)were higher than unilateral CI group, the statistical significances were seen at the 3rd, 6th, 12th, 18th and 24th month after switch-on(P<0.05). The MUSS scores of bimodal group after switch-on were higher than unilateral CI group(the 1st, 3rd, 6th, 12th, 18th and 24th month), the statistical significances were found at the 12th, 18th and 24th month, respectively(P<0.05). The SIR scores of bimodal group after switch-on were higher than unilateral CI group(the 1st, 3rd, 6th, 12th, 18th and 24th month), and significant differences appeared at the 12th, 18th and 24th month after switch-on(P<0.05).

Conclusion: Bimodal intervention could be helpful to the development of auditory and speech ability of infants.


BACKGROUND: Mutations in GJB3 were originally shown to underlie an autosomal dominant form of non-syndromic deafness in Chinese patients and the c.538C>T (p.R180*) variants caused high-frequency hearing loss. But after that, few reports have reported this mutation. This study investigated the relationship between the GJB3 c.538C>T variant and hearing phenotype in Chinese to assist with risk assessment and genetic counseling for hearing loss patients and their families.

METHOD: The study enrolled 5700 patients with hearing loss and 4600 normal subjects. Deafness gene mutations were distinguished using a gene chip. The GJB3 c.538C>T variant rate was calculated from the results. RESULT: Of the 5700 patients, 23 (0.40%) carried a GJB3 c.538C>T heterozygous variant; of these, 11 patients had other gene (GJB2/SLC26A4) mutations simultaneously. Most patients had moderate to profound hearing loss. All 23 patients were sporadic cases and had no family history of deafness. Of the 4600 normal individuals, 11 (0.24%) had GJB3 c.538C>T heterozygous variant. There was no statistical difference in incidence between the two groups.

CONCLUSIONS: Our results showed that the GJB3 c.538C>T variant has a very low incidence in the Chinese population, and there was no clear evidence to support a role of the GJB3 c.538C>T variant in the autosomal dominant form of non-syndromic deafness. Our findings suggested that GJB3 c.538C>T does not contribute to hearing loss, and this conclusion will assist with genetic counseling and risk prediction for deafness related to the GJB3 c.538C>T variant.

Johnson LC, Toro M, Vishnja E, Berish A, Mills B, Lu Z, Lieberman E.


BACKGROUND: Although the utility of universal newborn hearing screening is undisputed, testing protocols vary. In particular, the impact of the infant’s age at the time of automated auditory brainstem response (AABR) screening has not been well studied.

METHODS: We conducted a retrospective review of newborn hearing screening data in 6817 low-risk, term and late-preterm newborns at our large, urban, academic medical center for a 1-year period to analyze the impact of age and other factors on the screening failure rate and referral for diagnostic testing.

RESULTS: AABR screening failure rates decreased with postnatal age over the first 48 hours; 13.3% failed at <24 hours versus 3.8% at ≥48 hours (P < .0001). Infants who were initially tested at ≥36 hours failed repeat testing more often than those who were tested at <36 hours (11.5% vs 18.9%; P = .03). Other factors that were associated with failure included being a boy and of a race other than white. Sensorineural hearing loss (SNHL) was diagnosed in 18.6% of infants who failed their final screening at ≥48 hours compared with 2.8% of those whose final screening occurred earlier (P = .03). SNHL was more likely in infants who failed their first screening bilaterally (21.2%) than unilaterally (4.4%); P = .03.

CONCLUSIONS: Among healthy newborns, delaying AABR screening in the first 48 hours minimized failure rates. SNHL was 6 times as likely in infants who failed their final screening at ≥48 hours compared with those who were screened at
<48 hours of age. In our study, we offer guidance for nursery directors and audiologists who determine hearing screening protocols and counsel families about results.

**Conflict of interest statement:** POTENTIAL CONFLICT OF INTEREST: The authors have indicated they have no potential conflicts of interest to disclose.

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

**Parental knowledge and attitudes to childhood hearing loss and hearing services in the Solomon Islands.**


**OBJECTIVE:** An understanding of parental knowledge and attitudes towards childhood hearing loss is essential to the successful implementation of audiology services. The present study aimed to investigate parental knowledge and attitudes among parents in the Solomon Islands.

**METHODS AND MATERIALS:** A total of 100 mothers and 50 fathers were administered a questionnaire via semi-structured interviews.

**RESULTS:** Highest parental awareness of aetiology of childhood hearing loss was noted for otitis media (94%), noise exposure (87.3%), and family history (72.7%). The highest parental awareness concerning public health initiatives to reduce/prevent otitis media was noted for routine childhood immunizations (84%) and breast-feeding (76%). Higher rates of knowledge in fathers than in mothers included otitis media ($p = 0.038$), noise exposure ($p = 0.007$), and breast-feeding ($p = 0.031$). Approximately half of parents (56%) agreed that curses may cause hearing loss. Overall parental responses showed positive support for infant hearing screening programs (96%) and school-based ear and hearing health examinations (99.3%).

**CONCLUSIONS:** High levels of parental readiness and support for childhood hearing services in the Solomon Islands was evident. Knowledge of aetiology of childhood hearing loss was highest for otitis media, noise exposure, and family history. Knowledge and attitudes of fathers to childhood hearing loss and hearing services was either the same or better than that of mothers.

Keilmann A, Friese B, Lässig A, Hoffmann V.

**Validation of the second version of the LittlEARS® Early Speech Production Questionnaire (LEESPQ) in German-speaking children with normal hearing.**


**INTRODUCTION:** The introduction of neonatal hearing screening and the increasingly early age at which children can receive a cochlear implant has intensified the need for a validated questionnaire to assess the speech production of children aged 0-18. Such a questionnaire has been created, the LittlEARS® Early Speech Production Questionnaire (LEESPQ). This study aimed to validate a second, revised edition of the LEESPQ.

**METHODS AND MATERIALS:** Questionnaires were returned for 362 children with normal hearing. Completed questionnaires were analysed to determine if the LEESPQ is reliable, prognostically accurate, internally consistent, and if gender or multilingualism affects total scores.

**RESULTS:** Total scores correlated positively with age. The LEESPQ is reliable, accurate, and consistent, and independent of gender or lingual status. A norm curve was created. **DISCUSSION:** This second version of the LEESPQ is a valid tool to assess the speech production development of children with normal hearing, aged 0-18, regardless of their gender. As such, the LEESPQ may be a useful tool to monitor the development of paediatric hearing device users.

**CONCLUSION:** The second version of the LEESPQ is a valid instrument for assessing early speech production of children aged 0-18 months.

Kim SY, Choi BY, Jung EY, Park H, Yoo HN, Park KH.

**Risk factors for failure in the newborn hearing screen test in very preterm twins.**


**BACKGROUND:** We aimed to identify prenatal and postnatal risk factors associated with abnormal newborn hearing screen (NHS) results and subsequently confirmed sensorineural hearing loss (SNHL) in preterm twin neonates.

**METHODS:** Electronic medical records of 159 twin neonates who were born alive after ≤32 weeks were retrospectively reviewed for hearing loss in both ears. Histopathologic examination of the placenta was performed and clinical data, including method of conception and factors specific to twins, were retrieved from a computerized perinatal database. The main outcome measure was failure to pass the NHS test. The generalized estimation equations model was used for twins.

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


OBJECTIVE: The aim of this study was to discover Turkish regional differences in the risk factors of newborn hearing loss.

METHOD: A multi-centered retrospective design was used. A total of 443 children, registered to the national newborn hearing screening programme, with bilateral hearing loss, from five different regions of Turkey, were evaluated in terms of the types of hearing loss, the degree of hearing loss, the types of risk factors, parental consanguinity, age at diagnosis and age of auditory intervention, respectively.

RESULTS: There was no significant difference in the prevalence of hearing loss between regions ($\chi^2 = 3.210$, $P = 0.523$). Symmetric Sensorineural Hearing Loss (SSHL) was the most common type of HL in all regions (91.8%). Profound HL was the most common degree of HL in all regions (46.2%). There were statistically significant differences between regions in terms of types of HL ($\chi^2 = 14.151$, $P = 0.000$). At a total, 323 (72.9%) of subjects did not have any risk factors. There were statistically significant differences between regions in terms of the types of risk factors (pre, peri and post-natal) for SSNHL ($\chi^2 = 16.095$, $P = 0.000$). For all regions, the age of diagnosis was convenient with the JCIH criteria. However the age of hearing aid application was prolonged in some regions. There were statistically significant differences between regions in terms of the age of diagnosis ($\chi^2 = 93.570$, $P = 0.000$) and the age of auditory intervention ($\chi^2 = 47.323$, $P = 0.000$). The confounding effects of gender, age of diagnosis, age of hearing aids applications, HL in the family, types of risk factors for HL on SSNHL were detected. CONCLUSION: To reach the goal of a high quality newborn hearing screening, there is a need to develop an evidence-based standard for follow up guideline. In addition, risk factors should be re-evaluated according to regional differences and all regions should take their own precautions according to their evidence based data.


OBJECTIVES: This study aimed to objectively evaluate access to soft sounds (55 dB SPL) in paediatric CI users, all wearing MED-EL (Innsbruck, Austria) devices who were fitted with the objective electrically elicited stapedius reflex threshold (eSRT) fitting method, to track their cortical auditory evoked potential (CAEP) presence and latency, and to compare their CAEPs to those of normal-hearing peers.

METHODS: Forty-five unilaterally implanted, pre-lingually deafened MED-EL CI users, aged 12-48 months, underwent CAEP testing in the clinic at regular monthly intervals post switch-on. CAEPs were recorded in response to short speech tokens /m/, /g/ and /t/ presented in the free field at 55 dB SPL. Twenty children with normal hearing (NH), similarly aged, underwent CAEP testing once.

RESULTS: The proportion of present CAEPs increased and CAEP P1 latencies reduced significantly with post-implantation duration. CAEPs were scored based on their presence and age-appropriate P1 latency. These CAEP scores increased significantly with post-implantation duration. CAEP scores were significantly worse for the /m/ speech token compared to the other two tokens. Compared to the NH group, CAEP scores were significantly smaller for all post-implantation test intervals.

CONCLUSIONS: This study provides clinicians with a first step towards typical ranges of CAEP presence, latency, and derived CAEP score over the first months of MED-EL CI use. CAEPs within these typical ranges could validate intervention whereas less than optimum CAEPs could prompt clinicians to seek solutions in a timely manner. CAEPs could clinically validate whether a CI provides adequate access to soft sounds. This approach could form an alternative to behavioural soft sound access verification.


OBJECTIVES: To facilitate early diagnosis of infants with hearing loss, a universal newborn hearing screening program (UNHS) has been implemented in Hong Kong’s public hospitals for over a decade. However, there have been no known studies investigating parent attitudes to, and satisfaction with, UNHS since its launch in Hong Kong. The present study aimed to investigate knowledge of UNHS as well as infant hearing development, and attitudes and satisfaction with UNHS, in Hong Kong mothers with newborns. The study was designed to help evaluate and improve an established UNHS public hospital program, based on the perspectives of service users.

METHODS: A researcher-developed questionnaire was administered to 102 mothers whose newborn had received UNHS in the postnatal wards of a large public hospital in Hong Kong. The questionnaire considered parental knowledge
of UNHS and infant hearing development, attitudes and satisfaction toward public hospital UNHS. In the knowledge dimension, parents’ preferred time and location for pre-test information delivery, interpretation of screening results, and knowledge of hearing developmental milestones were surveyed. In addition, maternal attitudes to and satisfaction with UNHS screening services, the potential impact of UNHS on parent emotions and parent-baby bonding, attitudes toward informed consent, and willingness to comply with diagnostic assessment referral were also be surveyed.

**RESULTS:** Mean participant scores on knowledge of infant hearing development were relatively low (M = 2.59/6.0, SD = 0.90). Many mothers also underestimated the potential ongoing risks of hearing impairment in babies. Around 80% of mothers thought an infant could not have hearing impairment after passing the screening. In addition, one-third of mothers thought a baby could not later develop hearing impairment in infancy or childhood. In terms of attitudes and satisfaction, participants gave somewhat negative ratings for questions regarding receiving sufficient information about the screening (M = 2.90/5.0, SD = 1.27), screening procedure (M = 2.20/5.0, SD = 1.08), and sufficiency of information about results (M = 2.87/5.0, SD = 1.14). Nonetheless, participants gave positive ratings concerning whether screening could lead to early diagnosis (M = 4.61/5.0, SD = 0.57) and over 95% of mothers supported UNHS despite potential for false positive results. Mothers reported a high willingness to bring their baby to follow-up assessments if required (M = 4.53/5.0, SD = 0.56). Participants gave positive ratings for their level of satisfaction with the time and location of first UNHS information provision (M = 4.34/5.0, SD = 0.80) and the way permission was asked for screening the baby (M = 4.04/5.0, SD = 0.97) but alternative procedures were also recommended. Most recommendations focused on providing more information about the test and a more detailed explanation of screening results.

**CONCLUSIONS:** The survey results highlighted the need to provide more information to parents about infant hearing development to support home monitoring for signs of hearing loss after UNHS, as well as more detailed explanation and information regarding hearing screening and the implications of results to parents. Regardless of location, surveys of this type may provide valuable support for UNHS program quality assurance.

**Lanzieri TM, Chung W, Leung J, Caviness AC, Baumgardner JL, Blum P, Bialek SR, Demmler-Harrison G; Congenital Cytomegalovirus Longitudinal Study Group.**

**Hearing Trajectory in Children with Congenital Cytomegalovirus Infection.**


**Objectives** To compare hearing trajectories among children with symptomatic and asymptomatic congenital cytomegalovirus infection through age 18 years and to identify brain abnormalities associated with sensorineural hearing loss (SNHL) in asymptomatic case patients.

**Study Design** Longitudinal prospective cohort study.

**Setting** Tertiary medical center.

**Subjects and Methods** The study included 96 case patients (4 symptomatic and 92 asymptomatic) identified through hospital-based newborn cytomegalovirus screening from 1982 to 1992 and 72 symptomatic case patients identified through referrals from 1993 to 2005. We used growth curve modeling to analyze hearing thresholds (0.5-8 kHz) by ear with increasing age and Cox regression to determine abnormal findings on head computed tomography scan associated with SNHL (hearing threshold ≥25 dB in any audiometric frequency) among asymptomatic case patients.

**Results** Fifty-six (74%) symptomatic and 20 (22%) asymptomatic case patients had SNHL: congenital/early-onset SNHL was diagnosed in 78 (51%) and 10 (5%) ears, respectively, and delayed-onset SNHL in 25 (17%) and 20 (11%) ears; 49 (32%) and 154 (84%) ears had normal hearing. In affected ears, all frequency-specific hearing thresholds worsened with age. Congenital/early-onset SNHL was significantly worse (severe-profound range, >70 dB) than delayed-onset SNHL (mild-moderate range, 26-55 db). Frequency-specific hearing thresholds were significantly different between symptomatic and asymptomatic case patients at 0.5 to 1 kHz but not at higher frequencies (2-8 kHz). Among asymptomatic case patients, white matter lucency was significantly associated with SNHL by age 5 years (hazard ratio, 4.4; 95% CI, 1.3-15.6).

**Conclusion** Congenital/early-onset SNHL frequently resulted in severe to profound loss in symptomatic and asymptomatic case patients. White matter lucency in asymptomatic case patients was significantly associated with SNHL by age 5 years.

**Lim CH, Lim JH, Kim D, Choi HS, Lee DH, Kim DK.**

**Bony cochlear nerve canal stenosis in pediatric unilateral sensorineural hearing loss.**


**OBJECTIVES:** This study was performed to evaluate the frequency of bony cochlear nerve canal (BCNC) stenosis and its clinical significance in pediatric patients with unilateral sensorineural hearing loss (SNHL) of unknown etiolo.gy.

**MATERIALS AND METHODS:** We analyzed the medical records and temporal bone computed tomography (CT) results of patients less than 13 years of age with a diagnosis of unilateral SNHL of unknown etiology between July 2007 and July 2017. We compared the BCNC diameter between both sides and analyzed the age at diagnosis, degree of hearing loss, and accompanying inner ear anomalies.

**RESULTS:** In 42 patients, the mean age at diagnosis was 7.4 ± 3.6 years, and the average hearing level in the affected ear was 87.9 ± 20.0 dB HL (decibels hearing level). The average diameter of the BCNC was 1.22 ± 0.75 mm on the
affected side and 1.96 ± 0.52 mm on the normal side. The most suitable criterion for BCNC stenosis appeared to be a diameter of 1.2 mm by the recursive partitioning procedure. With application of this criterion, the rate of BCNC stenosis was significantly greater on the affected side than on the normal side (52.4% vs. 4.8%, respectively; \( P < 0.05 \)). A narrow internal acoustic canal was found in two patients, and vestibular and cochlear anomalies were found in three patients each.

**CONCLUSIONS:** Our results suggest that it is reasonable to set a diameter of 1.2 mm as a cutoff for BCNC stenosis, and also that BCNC stenosis is a common cause of unilateral SNHL of unknown etiology in childhood.


**OBJECTIVES:** To examine intelligence, language, and academic achievement through 18 years of age among children with congenital cytomegalovirus infection identified through hospital-based newborn screening who were asymptomatic at birth compared with uninfected infants.

**METHODS:** We used growth curve modeling to analyze trends in IQ (full-scale, verbal, and nonverbal intelligence), receptive and expressive vocabulary, and academic achievement in math and reading. Separate models were fit for each outcome, modeling the change in overall scores with increasing age for patients with normal hearing (\( n = 78 \)) or with sensorineural hearing loss (SNHL) diagnosed by 2 years of age (\( n = 11 \)) and controls (\( n = 40 \)).

**RESULTS:** Patients with SNHL had full-scale intelligence and receptive vocabulary scores that were 7.0 and 13.1 points lower, respectively, compared with controls, but no significant differences were noted in these scores among patients with normal hearing and controls. No significant differences were noted in scores for verbal and nonverbal intelligence, expressive vocabulary, and academic achievement in math and reading among patients with normal hearing or with SNHL and controls.

**CONCLUSIONS:** Infants with asymptomatic congenital cytomegalovirus infection identified through newborn screening with normal hearing by age 2 years do not appear to have differences in IQ, vocabulary or academic achievement scores during childhood, or adolescence compared with uninfected children.

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Conflict of interest statement: POTENTIAL CONFLICT OF INTEREST: Dr Demmler-Harrison’s institution has received funding from Merck Sharpe & Dohme Corporation since July 2016 to assist with salary support for further analysis on long-term outcomes of congenital cytomegalovirus infection not included in this report; the other authors have indicated they have no potential conflicts of interest to disclose.

Lu X, Qin Z.
Auditory and language development in Mandarin-speaking children after cochlear implantation.

**OBJECTIVE:** To evaluate early auditory performance, speech perception and language skills in Mandarin-speaking pre-lingual deaf children in the first two years after they received a cochlear implant (CI) and analyse the effects of possible associated factors.

**METHODS:** The Infant-Toddler Meaningful Auditory Integration Scale (ITMAIS)/Meaningful Auditory Integration Scale (MAIS), Mandarin Early Speech Perception (MESP) test and Putonghua Communicative Development Inventory (PCDI) were used to assess auditory and language outcomes in 132 Mandarin-speaking children pre- and post-implantation.

**RESULTS:** Children with CIs exhibited an ITMAIS/MAIS and PCDI developmental trajectory similar to that of children with normal hearing. The increased number of participants who achieved MESP categories 1-6 at each test interval showed a significant improvement in speech perception by paediatric CI recipients. Age at implantation and socioeconomic status were consistently associated with both auditory and language outcomes in the first two years post-implantation.

**CONCLUSION:** Mandarin-speaking children with CIs exhibit significant improvements in early auditory and language development. Though these improvements followed the normative developmental trajectories, they still exhibited a gap compared with normative values. Earlier implantation and higher socioeconomic status are consistent predictors of greater auditory and language skills in the early stage.

Progressive deafness-dystonia due to SERAC1 mutations: A study of 67 cases. 

OBJECTIVE: 3-Methylglutaconic aciduria, dystonia-deafness, hepatopathy, encephalopathy, Leigh-like syndrome (MEG-DHEL) syndrome is caused by biallelic variants in SERAC1.

METHODS: This multicenter study addressed the course of disease for each organ system. Metabolic, neuroradiological, and genetic findings are reported.

RESULTS: Sixty-seven individuals (39 previously unreported) from 59 families were included (age range = 5 days-33.4 years, median age = 9 years). A total of 41 different SERAC1 variants were identified, including 20 that have not been reported before. With the exception of 2 families with a milder phenotype, all affected individuals showed a strikingly homogeneous phenotype and time course. Severe, reversible neonatal liver dysfunction and hypoglycemia were seen in >40% of all cases. Starting at a median age of 6 months, muscular hypotonia (91%) was seen, followed by progressive spasticity (82%, median onset = 15 months) and dystonia (82%, 18 months). The majority of affected individuals never learned to walk (68%). Seventy-nine percent suffered hearing loss, 58% never learned to speak, and nearly all had significant intellectual disability (88%). Magnetic resonance imaging features were accordingly homogenous, with bilateral basal ganglia involvement (98%); the characteristic “putaminal eye” was seen in 53%. The urinary marker 3-methylglutaconic aciduria was present in virtually all patients (98%). Supportive treatment focused on spasticity and drooling, and was effective in the individuals treated; hearing aids or cochlear implants did not improve communication skills.

INTERpretation: MEGDHEL syndrome is a progressive deafness-dystonia syndrome with frequent and reversible neonatal liver involvement and a strikingly homogenous course of disease. Ann Neurol 2017;82:1004-1015

Mallen JR, Hunter JB, Auerbach C, Wexler L, Vambutas A.
Characterization of newborn hearing screening failures in multigestational births.

OBJECTIVE: To define the rate and characterize the type of newborn hearing screening failures in multigestational births.

METHODS: Retrospective chart review of all multigestational births that occurred in a 10-year period (2002-2012) in which at least one newborn failed newborn hearing screening at two tertiary care hospitals in the Northwell Health System.

RESULTS: Out of 125,405 total births, we identified 2961 multigestational births, of which 59 (2.0%) newborns failed newborn hearing screening. None of their 66 twin/triplet siblings failed their newborn hearing screens. Of 43 newborns that returned for follow-up, 56.0% (24/43) had confirmed hearing loss, resulting in an overall rate of 0.81% in all multigestational newborns with hearing loss. Of 19 infants that passed repeat testing, two were judged to need myringotomy tube placement. Twenty-four infants had a confirmed hearing loss, 11 of which had sensorineural hearing loss (0.37%), and 13 with a conductive or mixed hearing loss (0.44%).

CONCLUSIONS: We identified a greater than expected risk of conductive hearing loss, not attributable to otitis media, than sensorineural hearing loss in this population. These observations are consistent with the increased risk of birth defects in multigestational births.

Matsushima K, Nakano A, Arimoto Y, Mutai H, Yamazawa K, Murayama K, Matsunaga T.
High-level heteroplasmy for the m.7445A>G mitochondrial DNA mutation can cause progressive sensorineural hearing loss in infancy.

OBJECTIVE: Hearing loss caused by mutation of mitochondrial DNA typically develops in late childhood or early adulthood, but rarely in infancy. We report the investigation of a patient to determine the cause of his early onset hearing loss.

MATERIALS AND METHODS: The proband was a boy aged 1 year and 2 months at presentation. Newborn hearing screening test by automated auditory brainstem response generated “pass” results for both ears. His reaction to sound deteriorated by 9 months. Average pure tone threshold at 0.5, 1, and 2 kHz was 55 dB by conditioned orientation audiometry. His father had congenital hearing loss, and his mother had progressive hearing loss since childhood. Invader assays and Sanger sequencing were performed to investigate genetic causes of the hearing loss in the proband, and heteroplasmy was assessed by PCR-restriction fragment length polymorphism, Sanger sequencing, and pyrosequencing. Additionally, mitochondrial function was evaluated by measurement of the oxygen consumption rate of patient skin fibroblasts.

RESULTS: An m.7445A > G mitochondrial DNA mutation and a heterozygous c.235delC (p.L79Cfs*3) mutation of GJB2 were detected in the proband. His mother carried the m.7445A > G mitochondrial DNA mutation, and his father was a compound heterozygote for GJB2 mutations (c.235delC; [134G > A; 408C > A]). Tissue samples from both the proband and his mother exhibited a high degree of heteroplasmy. Fibroblasts from the proband exhibited markedly reduced oxygen consumption rates. These data indicate that the proband had impaired mitochondrial function, resulting in hearing loss.

CONCLUSION: This research demonstrates that hearing loss in a proband who presented in infancy and that of his mother resulted from a high level of heteroplasmy for the m.7445A > G mitochondrial DNA mutation, indicating that this alteration can cause hearing loss in infancy.
Hyperacusis in children: A clinical profile.

Myne S, Kennedy V.


OBJECTIVE: Hyperacusis is commonly seen in clinical paediatric practice and can be distressing for the children and their families. This paper looks at the clinical profile of children seen for hyperacusis in a paediatric audiology service and reviews the possible underlying mechanisms.

METHODS: Retrospective study of case notes of 61 children with troublesome hyperacusis seen in the paediatric audiology service, looking at their clinical presentation and presence of other medical conditions.

RESULTS: Hyperacusis was the main presenting complaint in more than half of the cases (n = 31, 51%). The commonest age at presentation with this problem was 3-4 years (n = 33, 54%). Hearing was normal in the majority of these children (n = 41, 67%). An active middle ear problem was observed in nearly half (n = 29, 48%) of all the children, of which glue ear (otitis media with effusion, OME) was the commonest. Presence of a neurodevelopmental condition was found in almost half (n = 28, 46%) of these patients of which autistic spectrum disorder was the commonest (8/61, 13%). In nearly one-fourth of the children (23%), presence of both middle ear problems and neurodevelopmental was noted. Tinnitus was an accompanying symptom reported in 11% of all the patients.

CONCLUSION: Hyperacusis may commonly present at a very young age. Awareness of different clinical presentations, presence of other medical conditions and possible underlying pathomechanisms in children with hyperacusis can be helpful for clinicians in informing prognosis, counselling and in individualising management plan.
Norrix LW, Velenovsky D.

**Purpose:** The auditory brainstem response (ABR) is a powerful tool for making clinical decisions about the presence, degree, and type of hearing loss in individuals in whom behavioral hearing thresholds cannot be obtained or are not reliable. Although the test is objective, interpretation of the results is subjective.

**Method:** This review provides information about evidence-based criteria, suggested by the 2013 Newborn Hearing Screening Program guidelines, and the use of cross-check methods for making valid interpretations about hearing status from ABR recordings.

**Results:** The use of an appropriate display scale setting, templates of expected response properties, and objective criteria to estimate the appropriate noise, signal level, and signal-to-noise ratio will provide quality data for determining ABR thresholds. Cross-checks (e.g., immittance measures, otoacoustic emissions testing, functional indications of a child's hearing) are also needed to accurately interpret the ABR.

**Conclusions:** Using evidence-based ABR signal detection criteria and considering the results within the context of other physiologic tests and assessments of hearing function will improve the clinician's accuracy for detecting hearing loss and, when present, the degree of hearing loss. Diagnostic accuracy will ensure that appropriate remediation is initiated and that children or infants with normal hearing are not subjected to unnecessary intervention.

Olivera LS, Didoné DD, Durante AS.
Automated cortical auditory evoked potentials threshold estimation in neonates.

**INTRODUCTION:** The evaluation of Cortical Auditory Evoked Potential has been the focus of scientific studies in infants. Some authors have reported that automated response detection is effective in exploring these potentials in infants, but few have reported their efficacy in the search for thresholds.

**OBJECTIVE:** To analyze the latency, amplitude and thresholds of Cortical Auditory Evoked Potential using an automatic response detection device in a neonatal population.

**METHODS:** This is a cross-sectional, observational study. Cortical Auditory Evoked Potentials were recorded in response to pure-tone stimuli of the frequencies 500, 1000, 2000 and 4000Hz presented in an intensity range between 0 and 80dB HL using a single channel recording. P1 was performed in an exclusively automated fashion, using Hotelling’s T2 statistical test. The latency and amplitude were obtained manually by three examiners. The study comprised 39 neonates up to 28 days old of both sexes with presence of otoacoustic emissions and no risk factors for hearing loss.

**RESULTS:** With the protocol used, Cortical Auditory Evoked Potential responses were detected in all subjects at high intensity and thresholds. The mean thresholds were 24.8±10.4dB NA, 25±9.0dB NA, 28±7.8dB NA and 29.4±6.6dB HL for 500, 1000, 2000 and 4000Hz, respectively.

**CONCLUSION:** Reliable responses were obtained in the assessment of cortical auditory potentials in the neonates assessed with a device for automatic response detection.

Poonual W, Navacharoen N, Kangsanarak J, Namwongprom S, Saokaew S.
Hearing loss screening tool (COBRA score) for newborns in primary care setting.

**Purpose:** To develop and evaluate a simple screening tool to assess hearing loss in newborns. A derived score was compared with the standard clinical practice tool.

**Methods:** This cohort study was designed to screen the hearing of newborns using transiently evoked otoacoustic emission and auditory brain stem response, and to determine the risk factors associated with hearing loss of newborns in 3 tertiary hospitals in Northern Thailand. Data were prospectively collected from November 1, 2010 to May 31, 2012. To develop the risk score, clinical-risk indicators were measured by Poisson risk regression. The regression coefficients were transformed into item scores dividing each regression-coefficient with the smallest coefficient in the model, rounding the number to its nearest integer, and adding up to a total score.

**Results:** Five clinical risk factors (Craniofacial anomaly, Ototoxicity, Birth weight, family history [Relative] of congenital sensorineural hearing loss, and Apgar score) were included in our COBRA score. The screening tool detected, by area under the receiver operating characteristic curve, more than 80% of existing hearing loss. The positive-likelihood ratio of hearing loss in patients with scores of 4, 6, and 8 were 25.21 (95% confidence interval [CI], 14.69-43.26), 58.52 (95% CI, 36.26-94.44), and 51.56 (95% CI, 33.74-78.82), respectively. This result was similar to the standard tool (The Joint Committee on Infant Hearing) of 26.72 (95% CI, 20.59-34.66). **Conclusion:** A simple screening tool of five predictors provides good prediction indices for newborn hearing loss, which may motivate parents to bring children for further appropriate testing and investigations.

**Conflict of interest statement:** Conflicts of interest: No potential conflict of interest relevant to this article was reported.
The Burden of Congenital Cytomegalovirus Infection: A Prospective Cohort Study of 20 000 Infants in Finland.  

Background: Congenital cytomegalovirus (cCMV) infection is the most common congenital infection and causes significant morbidity. This study was undertaken to evaluate the benefits of screening newborns for cCMV and to understand the cCMV disease burden in Finland.

Methods: Infants born in Helsinki area hospitals were screened for CMV by testing their saliva with a real-time polymerase chain reaction assay. The CMV-positive infants and matched controls were monitored to determine their neurodevelopmental, audiological, and ophthalmological outcomes at 18 months of age. Griffiths Mental Development Scales, otoacoustic emission and sound field audiometry, and ophthalmologic examination were performed.

Results: Of the 19868 infants screened, 40 had confirmed cCMV infection (prevalence, 2 in 1000 [95% confidence interval, 1.4-2.6 in 1000]). Four (10%) infants had symptomatic cCMV. Griffiths general quotients did not differ significantly between the CMV-positive (mean, 101.0) and control (mean, 101.6) infants (P = .557), nor did quotients for any of the Griffiths subscales (locomotion, personal-social, hearing and language, eye and hand, performance) (P = .173-.721). Four of 54 CMV-positive ears and 6 of 80 CMV-negative ears failed otoacoustic emission testing (P = 1.000). The mean minimal response levels over the frequencies 500 Hz to 4 kHz in the sound field audiometry did not differ between CMV-positive (mean, 34.31-dB hearing level) and control (mean, 32.73-dB hearing level) infants (P = .338). No CMV-related ophthalmologic findings were observed.

Conclusions: The prevalence of cCMV was low, and outcomes at 18 months of age did not differ between the infected infants and healthy control infants. With such a low burden in Finland, universal newborn screening for cCMV seems unwarranted.

Pérez-Martín J, Artaso MA, Díez FJ.  
Cost-effectiveness of pediatric bilateral cochlear implantation in Spain.  

OBJECTIVES/HYPOTHESIS: To determine the incremental cost-effectiveness of bilateral versus unilateral cochlear implantation for 1-year-old children suffering from bilateral sensorineural severe to profound hearing loss from the perspective of the Spanish public health system.

STUDY DESIGN: Cost-utility analysis.

METHODS: We conducted a general-population survey to estimate the quality-of-life increase contributed by the second implant. We built a Markov influence diagram and evaluated it for a life-long time horizon with a 3% discount rate in the base case.

RESULTS: The incremental cost-effectiveness ratio of simultaneous bilateral implantation with respect to unilateral implantation for 1-year-old children with severe to profound deafness is €10,323 per quality-adjusted life year (QALY). For sequential bilateral implantation, it rises to €11,733/QALY. Both options are cost-effective for the Spanish health system, whose willingness to pay is estimated at around €30,000/QALY. The probabilistic sensitivity analysis shows that the probability of bilateral implantation being cost-effective reaches 100% for that cost-effectiveness threshold.

CONCLUSIONS: Bilateral implantation is clearly cost-effective for the population considered. If possible, it should be done simultaneously (i.e., in one surgical operation), because it is as safe and effective as sequential implantation, and saves costs for the system and for users and their families. Sequential implantation is also cost-effective for children who have received the first implant recently, but it is difficult to determine when it ceases to be so because of the lack of detailed data. These results are specific for Spain, but the model can easily be adapted to other countries.


Ravi R, Gunjawate DR, Yerraguntla K, Rajashekhar B.  

Systematic review of knowledge of, attitudes towards, and practices for newborn hearing screening among healthcare professionals.

INTRODUCTION: The success of newborn hearing screening programs lies in the timely identification, diagnosis, and management of children with hearing loss accomplished via a multidisciplinary newborn hearing screening (NHS) team. The team is typically comprised of various healthcare professionals who act as decision makers as well as facilitators for different stages in the screening process. Team members’ knowledge of, attitudes towards, and practices for early hearing detection and intervention programs are critical for success and prevention of loss to follow up. In this context, it becomes crucial to understand their knowledge of, attitudes towards, and practices for towards newborn hearing screening.

METHODS: A systematic review was conducted on the following databases; PubMed/Medline, Cumulative Index to Nursing and Allied Health Literature (CINAHL), Scopus, Web of Science, Science Direct and Cochrane Library. This search was carried out using various keywords such as practitioners, newborn hearing screening, knowledge, attitudes, and practices in different combinations. The review was conducted based on Preferred Reporting Items for Systematic
RESULTS: A total of 271 hits were obtained of which 20 articles were found suitable for inclusion in the final review. Overall, similar results were found regarding team members’ knowledge of NHS programs, regardless of country of origin. Similarly, attitudes toward NHS programs were positive. Team members’ experiences with NHS programs varied from country-to-country and across healthcare professionals. Results consistently showed gaps in team members’ knowledge suggesting the need for outreach and professional education programs on NHS.

CONCLUSION: NHS teams members from different countries, healthcare systems, and early hearing detection and intervention programs show gaps in critical knowledge warranting outreach and educational programs.

Neonates with congenital Cytomegalovirus and hearing loss identified via the universal newborn hearing screening program.

BACKGROUND: Congenital cytomegalovirus (CMV) is the most common non-genetic cause of sensorineural hearing loss. Currently, there are no universal CMV screening programs for newborns or routine CMV testing of neonates with hearing loss in Australia, or elsewhere. OBJECTIVES: This study was undertaken to determine the prevalence of congenital CMV infection in infants with hearing loss identified using routine resources via the Australian universal neonatal hearing screening (UNHS) program.

STUDY DESIGN: Infants who failed UNHS, referred for audiological testing and found to have permanent hearing loss were screened for CMV via PCR of urine and saliva. Congenital CMV was diagnosed if CMV was detected in infants ≤30 days of age, or using retrospective testing on stored new born screening cards, retrospective testing, or using clinical criteria if >30 days of age. The cohort was analyzed for time of testing and prevalence of congenital CMV determined. RESULTS: The Audiology Department reviewed 1669 infants who failed UNHS between 2009 and 2016. Thirty percent (502/1669) had permanent hearing loss confirmed, of whom 336/502 were offered CMV testing. A definite (n = 11) or probable (n = 8) diagnosis of congenital CMV occurred in 19/323 (5.9%), of whom definite diagnoses were made in 4/19 on tests positive prior to 21 days of life, in 5/19 who were positive on neonatal blood screening card (NBSC) testing, in 2/19 who were positive on placental testing. In 8/19 probable diagnoses were made based on positive testing between ages 23-42 days and a consistent clinical syndrome in the absence of another cause for hearing loss after genetic and other testing. CMV testing mirrored the timing of audiological testing, with ~40% completing audiology and CMV testing by 21 days, and 64% by 30 days.

CONCLUSION: This program, utilizing existing clinical services identified probable congenital CMV in ~6% of a large cohort failing UNHS with permanent hearing loss, of whom more than half were definite diagnoses. No additional assets were required to those already existing in this tertiary referral pediatric centre, whilst providing useful and timely data for clinical and audiological management.

Razza S, Zaccone M, Meli A, Cristofari E. 
Evaluation of speech reception threshold in noise in young Cochlear® Nucleus® system 6 implant recipients using two different digital remote microphone technologies and a speech enhancement sound processing algorithm.

OBJECTIVE: Children affected by hearing loss can experience difficulties in challenging and noisy environments even when deafness is corrected by Cochlear implant (CI) devices. These patients have a selective attention deficit in multiple listening conditions. At present, the most effective ways to improve the performance of speech recognition in noise consists of providing CI processors with noise reduction algorithms and of providing patients with bilateral CIs. The aim of this study was to compare speech performances in noise, across increasing noise levels, in CI recipients using two kinds of wireless remote-microphone radio systems that use digital radio frequency transmission: the Roger Inspiro accessory and the Cochlear Wireless Mini Microphone accessory.

METHODS: Eleven Nucleus Cochlear CP910 CI young user subjects were studied. The signal/noise ratio, at a speech reception threshold (SRT) value of 50%, was measured in different conditions for each patient: with CI only, with the Roger or with the MiniMic accessory. The effect of the application of the SNR-noise reduction algorithm in each of these conditions was also assessed. The tests were performed with the subject positioned in front of the main speaker, at a distance of 2.5 m. Another two speakers were positioned at 3.50 m. The main speaker at 65 dB issued disyllabic words. Babble noise signal was delivered through the other speakers, with variable intensity.

RESULTS: The use of both wireless remote microphones improved the SRT results. Both systems improved gain of speech performances. The gain was higher with the Mini Mic system (SRT = -4.76) than the Roger system (SRT = -3.01). The addition of the NR algorithm did not statistically further improve the results.

CONCLUSION: There is significant improvement in speech recognition results with both wireless digital remote microphone accessories, in particular with the Mini Mic system when used with the CP910 processor. The use of a remote microphone accessory surpasses the benefit of application of NR algorithm.
Laryngeal mask airway (LMA) may be less stimulating to the airway and allow for shorter overall operating room time. Previous studies report LMA use during adenotonsillectomy with conversion rates to ETT of up to 17%. There has been no prior evaluation of LMA use during adenoidectomy alone. In this study, we attempt to identify the rate and contributing factors of LMA failure during adenoidectomy.

METHODS: All pediatric patients undergoing adenoidectomy between January 1, 2016 and June 30, 2017 were reviewed. Demographic and clinical data were collected and analysed to determine the need for conversion to ETT and the occurrence of any complications.

RESULTS: Our study revealed 139 pediatric patients who underwent adenoidectomy during the study period. 110 patients had adenoidectomy performed with LMA and 27 patients had ETT. Two patients (1.8%) required conversion to ETT because of difficulty with ventilation when the mouth gag was in place. There were no complications. Mean operating room time was 20 min less in the LMA group (P < 0.05).

CONCLUSIONS: The use of an LMA in adenoidectomy may be a safe and effective alternative to ETT. More study is required to determine overall complication rates.

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evidence of Zika virus infection were assessed through clinical evaluations, caregiver interviews, and review of medical records. At follow-up (ages 19-24 months), most of these children had severe motor impairment, seizure disorders, hearing and vision abnormalities, and sleep difficulties. Children with microcephaly and laboratory evidence of Zika virus infection have severe functional limitations and will require specialized care from clinicians and caregivers as they age.


OBJECTIVE: Temporary conductive hearing loss due to amniotic fluid accumulation in the middle ear cavity may lead to failure (false positive) in newborn hearing screening tests. The aim of this study was to identify whether amniotic fluid index has association with failure of the initial newborn otoacoustic emission (OAE) screening test.

METHODS: A cohort study in a tertiary hospital center (Royal Victoria Hospital, Montréal) was constructed from 70 newborns that failed the OAE test, but passed a subsequent auditory brainstem response (ABR) test, and 75 randomly selected newborns that passed initial otoacoustic emission testing. Maternal (including the amniotic fluid index in the third trimester) and newborn clinical data were extracted from medical records. Statistical association models were built to determine variables that influenced hearing screen passage or failure.

RESULTS: The two arms of the cohort had no significant differences in maternal or child clinical indices, including in amniotic fluid index. Calculated as individual odds ratios, maternal tobacco [95% CI of odds ratio: 0.04, 0.59, p = 0.0078], and drug use [95% CI of odds ratio: 0.0065, 0.72, p = 0.058] [borderline significance] were associated with failing the otoacoustic emission testing.

CONCLUSIONS: Amniotic fluid index was not found to be associated with failure of otoacoustic emission screening in newborns. However, our study unveiled an interesting unexpected association of OAE failure with maternal smoking and/or drug use. This finding can help alleviate some of the time, cost and parental anxiety related to failed OAE screening. In selected cases of maternal smoking or drug use we might want to replace or add OAE to the ABR test in newborn hearing screening protocols, that don’t perform both tests before discharge.


The Newborn Hearing Screening Programme (NHSP) was established in Cork University Maternity Hospital (CUMH) in April 2011. Between April 2011 and July 2014, 42 infants were identified with a Permanent Childhood Hearing Impairment (PCHI). Following this diagnosis, infants underwent a paediatric assessment according to recognised guidelines with the intention of identifying the underlying aetiology of the PCHI. The aim of this study was to assess the findings of this aetiological workup via retrospective chart review. PCHI data was obtained from the eSP database. This is a web based information system (eSP) used to track each baby through the screening and referral process A retrospective chart review of these patients was performed. Sixteen (38%) infants were diagnosed with a bilateral sensorineural hearing loss. Two infants had congenital CMV infection. A Connexin 26 gene mutation was detected in one infant. Five babies underwent cochlear implantation. Through adherence to the recommended protocol a possible cause of PCHI may be determined. This study has identified areas of future improvement for this service in Ireland.


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


OBJECTIVE: To evaluate the clinically relevant abnormalities as visualized on CT and MR imaging in children with symmetric and asymmetric bilateral sensorineural hearing loss (SNHL), in relation to age and the severity of hearing loss.
STUDY DESIGN: Retrospective cohort study.
SETTING: Tertiary referral otology and audiology center.
PATIENTS AND DIAGNOSTIC INTERVENTIONS: From January 2006 until January 2016, a total of 207 children diagnosed with symmetric and asymmetric bilateral SNHL were included. They underwent CT and/or MR imaging for the evaluation of the etiology of their hearing loss.
MAIN OUTCOME MEASURES: Radiologic abnormalities associated with SNHL.
RESULTS: 302 scans were performed in 207 children (median age of 0.8 years old) with bilateral SNHL. The most frequently identified cause of bilateral SNHL was a malformation of the labyrinth. The combined diagnostic yield of CT and MR imaging was 32%. The diagnostic yield of MR (34%) was considerably higher than that of CT (20%). We found a higher rate of abnormalities in children with profound hearing loss (41%) compared to milder hearing loss (8-29%), and in asymmetric SNHL (52%) compared to symmetric SNHL (30%).
CONCLUSION: Imaging is essential in the etiologic evaluation of children with bilateral SNHL. The highest diagnostic yield is found in children with bilateral asymmetric SNHL or profound SNHL. Based on our findings, MR is the primary imaging modality of choice in the etiological evaluation of children with bilateral SNHL because of its high diagnostic yield.

Vancor E, Shapiro ED, Loyal J. 
Results of a Targeted Screening Program for Congenital Cytomegalovirus Infection in Infants Who Fail Newborn Hearing Screening. 

Background: Congenital cytomegalovirus (CMV) infection is a major cause of sensorineural hearing loss. By law, newborns in Connecticut who fail newborn hearing screening are tested for infection with CMV. This targeted screening is controversial, because most children with congenital CMV infection are asymptomatic, and CMV-related hearing loss can have a delayed onset. Our hospital uses a saliva polymerase chain reaction (PCR) assay (confirmed by a urine PCR assay) to detect CMV. Here, we report the results of the first year of our screening program.

Methods: We reviewed the medical records of newborns in the Yale New Haven Health System who failed the newborn hearing screening test between January 1 and December 31, 2016.
Results: Of 10964 newborns, 171 failed newborn hearing screening, and 3 of these newborns had positive saliva CMV PCR test results. Of these 3 newborns, 2 had positive results on the confirmatory test (for 1 of them the confirmatory test was not performed until the infant was 10 weeks old), and 1 had a negative result on the confirmatory test. Three additional newborns with congenital CMV infection were tested because of clinical indications (1 for ventriculomegaly on prenatal ultrasound and 2 for CMV infection of the mother). Results of audiology follow-up were available for 149 (87.1%) of the 171 newborns who failed newborn hearing screening; 127 (85.2%) had normal results.
Conclusion: Our targeted screening program for congenital CMV infection had a low yield. Consideration should be given to other strategies for identifying children at risk of hearing loss as a result of congenital CMV infection.

Verkerk MM, Wagner R, Fishchuk R, Fagan JJ. 
Survey of otolaryngology services in Ukraine and neighbouring Central and Eastern European countries. 

OBJECTIVE: The present humanitarian crisis in Ukraine is putting strains on its healthcare system. This study aimed to assess services and training in otolaryngology, audiology and speech therapy in Ukraine and its geographical neighbours.

METHOD: Survey study of 327 otolaryngologists from 19 countries.
RESULTS: Fifty-six otolaryngologists (17 per cent) from 15 countries responded. Numbers of otolaryngologists varied from 3.6 to 12.3 per 100 000 population (Ukraine = 7.8). Numbers of audiologists varied from 0, in Ukraine, to 2.8 per 100 000, in Slovakia, and numbers of speech therapists varied from 0, in Bulgaria, to 4.0 per 100 000, in Slovenia (Ukraine = 0.1). Ukraine lacks newborn and school hearing screening, good availability of otological drills and microscopes, and a cochlear implant programme.
CONCLUSION: There is wide variation in otolaryngology services in Central and Eastern Europe. All countries surveyed had more otolaryngologists per capita than the UK, but availability of audiology and speech and language therapy is poor. Further research on otolaryngology health outcomes in the region will guide service improvement.

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

BACKGROUND: Hearing loss is a barrier to speech and social and cognitive development. This can be especially pronounced in children living in low- and middle-income countries with limited resources. AIM: To determine the feasibility, durability and social impact of ComCare GLW solar-powered hearing aids provided for Vietnamese children with hearing impairment.
CASE: Sonia is a 4 years 1 month-year-old girl with Waardenburg syndrome and bilateral sensorineural hearing loss who had bilateral cochlear implants at 2 years 7 months years of age. She is referred to Developmental-Behavioral Pediatrics by her speech/language pathologist because of concerns that her language skills are not progressing as expected after the cochlear implant. At the time of the implant, she communicated using approximately 20 signs and 1 spoken word (mama). At the time of the evaluation (18 months after the implant) she had approximately 70 spoken words (English and Spanish) and innumerable signs that she used to communicate. She could follow 1-step directions in English but had more difficulty after 2-step directions. Sonia was born in Puerto Rico at 40 weeks gestation after an uncomplicated pregnancy. She failed her newborn hearing test and was given hearing aids that did not seem to help. At age 2 years, Sonia, her mother, and younger sister moved to the United States where she was diagnosed with bilateral severe-to-profound hearing loss. Genetic testing led to a diagnosis of Waardenburg syndrome (group of genetic conditions that can cause hearing loss and changes in coloring [pigmentation] of the hair, skin, and eyes). She received bilateral cochlear implants 6 months later. Sonia’s mother is primarily Spanish-speaking and mostly communicates with her in Spanish or with gestures but has recently begun to learn American Sign Language (ASL). In a preschool program at a specialized school for the deaf, Sonia is learning both English and ASL. Sonia seems to prefer to use ASL to communicate.

RESULTS: Hearing aids were well tolerated for use during regular school hours. All units remained functional during the study period (12 months). Teachers noted increased student awareness and responsiveness to surrounding sounds, but the degree of response to amplification varied between children. There was no significant improvement in speech development as all subjects had prelingual deafness. Teachers felt confident in troubleshooting any potential device malfunction.

CONCLUSIONS: A solar-powered hearing aid may be a viable option for children in low- and middle-income countries. This study demonstrates that device distribution, maintenance and function can be established in countries with limited resources, while providing feasibility data to support future studies investigating how similar devices may improve the quality of life of those with hearing loss.


BACKGROUND: Newborn hearing screening programs aim to lower the ages at audiological intervention among hearing-impaired children. In Wallonia and Brussels (Belgium), audiological intervention data are not collected in the screening program, and the ages at initiating audiological care have never been assessed. This study aimed to assess the evolution in the ages at initiating audiological intervention in the context of a newborn hearing screening program implementation.

METHODS: This population-based descriptive study used data from the Belgian healthcare billing database. The main outcomes were the children’s ages at the initial audiological assessment, hearing-aid fitting, and cochlear implantation. Results were compared to the same outcomes from another Belgian regional program (Flanders) that was implemented one decade earlier. Annual birth cohorts from 2006 to 2011 were included in the study.

RESULTS: In Wallonia-Brussels, the median ages for all outcomes tended to decrease over time but remained higher than in Flanders for each birth cohort. For all outcomes except the hearing-aid fitting, differences in median ages between the two regions became less pronounced during the study period. In 2006, <23% of the children from Wallonia-Brussels received any audiological care before the age of 12 months and these proportions were approximately 2-fold greater in the subsequent birth cohorts. For all outcomes, early care (<12 months) was typically delivered less frequently in Wallonia-Brussels, compared to the delivery in Flanders. These region-specific differences exhibited a decreasing trend over time, and statistically significant differences were less common in the later birth cohorts.

CONCLUSIONS: We conclude that the hearing screening program in Wallonia and Brussels promoted earlier audiological intervention among hearing-impaired children. However, milestones recommended by experts for an early intervention were not totally encountered. We also recommend collecting audiological intervention data as part of this program, which can facilitate more accurate and regular program evaluation.


CASE: Sonia is a 4 years 1 month-year-old girl with Waardenburg syndrome and bilateral sensorineural hearing loss who had bilateral cochlear implants at 2 years 7 months years of age. She is referred to Developmental-Behavioral Pediatrics by her speech/language pathologist because of concerns that her language skills are not progressing as expected after the cochlear implant. At the time of the implant, she communicated using approximately 20 signs and 1 spoken word (mama). At the time of the evaluation (18 months after the implant) she had approximately 70 spoken words (English and Spanish) and innumerable signs that she used to communicate. She could follow 1-step directions in English but had more difficulty after 2-step directions. Sonia was born in Puerto Rico at 40 weeks gestation after an uncomplicated pregnancy. She failed her newborn hearing test and was given hearing aids that did not seem to help. At age 2 years, Sonia, her mother, and younger sister moved to the United States where she was diagnosed with bilateral severe-to-profound hearing loss. Genetic testing led to a diagnosis of Waardenburg syndrome (group of genetic conditions that can cause hearing loss and changes in coloring [pigmentation] of the hair, skin, and eyes). She received bilateral cochlear implants 6 months later. Sonia’s mother is primarily Spanish-speaking and mostly communicates with her in Spanish or with gestures but has recently begun to learn American Sign Language (ASL). In a preschool program at a specialized school for the deaf, Sonia is learning both English and ASL. Sonia seems to prefer to use ASL to communicate. Sonia receives speech and language therapy (SLT) 3 times per week (90 minutes total) individually in school and once per week within a group. She is also receiving outpatient SLT once per week. Therapy sessions are completed in English, with the aid of an ASL interpreter. Sonia’s language scores remain low, with her receptive skills in the first percentile, and her expressive skills in the fifth percentile. During her evaluation in Developmental and Behavioral Pediatrics, an ASL interpreter...
was present, and the examiner is a fluent Spanish speaker. Testing was completed through a combination of English, Spanish, and ASL. Sonia seemed to prefer ASL to communicate, although she used some English words with errors of pronunciation. On the Beery Visual-Motor Integration Test, she obtained a standard score of 95. Parent and teacher rating scales were not significant for symptoms of attention-deficit/hyperactivity disorder. What factors are contributing to her slow language acquisition and how would you modify her treatment plan?

Wang J, Sun J, Sun J, Chen J.

Variations in electrode impedance during and after cochlear implantation: Round window versus extended round window insertions.


**OBJECTIVES:** To assess differences in intra- and postoperative electrode impedances following cochlear implantation between round window insertions (RWI) and extended round window insertions (ERWI).

**METHODS:** Fifty patients with congenital hearing loss received unilateral hearing implants (Sonata Ti100, Med-El GmbH, Innsbruck, Austria) with standard electrode arrays. The patients were divided into two groups according to the surgical technique used. Thirty-five procedures were performed with RWI (group A) and 15 with ERWI (group B). Electrode impedance was measured and analysed during the operation, and one week and one month postoperatively.

**RESULTS:** There were no statistically significant differences (i.e., P > 0.05) in electrode impedance between groups A and B intraoperatively, or at one week or one month postoperatively. Electrode impedance at one month postoperatively was higher than the intraoperative and postoperative one week values in group A (P < 0.05), with similar results in group B.

**CONCLUSION:** There was no significant difference between RWI and ERWI in operative duration or complications of cochlear implantation. Moreover, no significant differences in postoperative electrode impedance values were found between the two surgical routes.

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

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


**OBJECTIVE:** The objective of this study was to explore the knowledge and attitude among mothers of newborns regarding infant hearing loss (HL) in Changsha, Hunan province, China.

**DESIGN:** A questionnaire including 18 items was given to mothers. STUDY SAMPLE: A total of 115 mothers participated in the study.

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

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


Hearing improvement with softband and implanted bone-anchored hearing devices and modified implantation surgery in patients with bilateral microtia-atresia.


**OBJECTIVE:** To evaluate auditory development and hearing improvement in patients with bilateral microtia-atresia using softband and implanted bone-anchored hearing devices and to modify the implantation surgery.

**METHODS:** The subjects were divided into two groups: the softband group (40 infants, 3 months to 2 years old, Ponto softband) and the implanted group (6 patients, 6-28 years old, Ponto). The Infant-Toddler Meaning Auditory Integration Scale was used conducted to evaluate auditory development at baseline and after 3, 6, 12, and 24 months, and visual reinforcement audiometry was used to assess the auditory threshold in the softband group. In the implanted group, bone-anchored hearing devices were implanted combined with the auricular reconstruction surgery, and high-resolution CT was used to assess the deformity preoperatively. Auditory threshold and speech discrimination scores of the patients with implants were measured under the unaided, softband, and implanted conditions.

**RESULTS:** Total Infant-Toddler Meaning Auditory Integration Scale scores in the softband group improved significantly and approached normal levels. The average visual reinforcement audiometry values under the unaided and softband conditions were 76.75 ± 6.05 dB HL and 32.25 ± 6.20 dB HL (P < 0.01), respectively. In the implanted group, the auditory thresholds under the unaided, softband, and implanted conditions were 59.17 ± 3.76 dB HL, 32.5 ± 2.74 dB HL, and 17.5 ± 5.24 dB HL (P < 0.01), respectively. The respective speech discrimination scores were 23.33 ± 14.72%, 77.17 ± 6.46%, and 96.50 ± 2.66% (P < 0.01).
CONCLUSIONS: Using softband bone-anchored hearing devices is effective for auditory development and hearing improvement in infants with bilateral microtia-atresia. Wearing softband bone-anchored hearing devices before auricle reconstruction and combining bone-anchored hearing device implantation with auricular reconstruction surgery may be the optimal clinical choice for these patients, and results in more significant hearing improvement and minimal surgical and anesthetic injury.

Wang Y, Bergeson TR, Houston DM.
Infant-Directed Speech Enhances Attention to Speech in Deaf Infants With Cochlear Implants.
Purpose: Both theoretical models of infant language acquisition and empirical studies posit important roles for attention to speech in early language development. However, deaf infants with cochlear implants (CIs) show reduced attention to speech as compared with their peers with normal hearing (NH; Horn, Davis, Pisoni, & Miyamoto, 2005; Houston, Pisoni, Kirk, Ying, & Miyamoto, 2003), which may affect their acquisition of spoken language. The main purpose of this study was to determine (a) whether infant-directed speech (IDS) enhances attention to speech in infants with CIs, as compared with adult-directed speech (ADS), and (b) whether the degree to which infants with CIs pay attention to IDS is associated with later language outcomes.

Method: We tested 46 infants-12 prelingually deaf infants who received CIs before 24 months of age and had 12 months of hearing experience (CI group), 22 hearing experience-matched infants with NH (NH-HEM group), and 12 chronological age-matched infants with NH (NH-CAM group)-on their listening preference in 3 randomized blocks: IDS versus silence, ADS versus silence, and IDS versus ADS. We administered the Preschool Language Scale-Fourth Edition (PLS-4; Zimmerman, Steinier, & Pond, 2002) approximately 18 months after implantation to assess receptive and expressive language skills of infants with CIs.

Results: In the IDS versus silence block, all 3 groups looked significantly longer to IDS than to silence. In the ADS versus silence block, both the NH-HEM and NH-CAM groups looked significantly longer to ADS relative to silence; however, the CI group did not show any preference. In the IDS versus ADS block, whereas both the CI and NH-HEM groups preferred IDS over ADS, the NH-CAM group looked equally long to IDS and ADS. IDS preference quotient among infants with CIs in the IDS versus ADS block was associated with PLS-4 Auditory Comprehension and PLS-4 Expressive Communication measures.

Conclusions: Two major findings emerge: (a) IDS enhances attention to speech in deaf infants with CIs; (b) the degree of IDS preference over ADS relates to language development in infants with CIs. These results support a focus on input in developing intervention strategies to mitigate the effects of hearing loss on language development in infants with hearing loss.

Characteristics of electrically evoked auditory brainstem responses in patients with cochlear nerve canal stenosis receiving cochlear implants.
OBJECTIVE: To explore the characteristics of the electrically evoked auditory brainstem responses (EABR) in children with cochlear nerve canal stenosis (CNCs) following cochlear implantation (CI), and the EABR thresholds in children with stenotic versus normal cochlear nerve canals.

METHOD: Sixteen children with profound sensorineural hearing loss were included in this study: 8 with CNCs (CNCs group) and 8 with normal cochlear nerve canals (control group). All children underwent cochlear implantation with full insertion of all electrodes. EABR was performed 6 months postoperatively in both groups.

RESULTS: The EABR extraction rate was 100% in children with normal cochlear nerve canals and only 50% in children with CNCs. EABR thresholds were significantly higher in children with CNCs of electrodes No. 11and 22 than in children with normal cochlear nerve canals (P < 0.05 for both comparisons). There was no significant difference in EABR thresholds among electrode No. 1, 11 and 22 in CNCs group (P > 0.05 for all comparisons); while in the control group, the EABR threshold at electrode No 22 was lower than those at both electrodes No. 11 and 1 (P < 0.05 for both comparisons), and the EABR threshold at electrode No. 11 was also lower than that at electrode No. 1 (P < 0.05).

CONCLUSION: The EABR thresholds in children with normal cochlear nerve canals vary according to the different locations of electrodes in the cochlea; while in children with CNCs, there was no significant difference among different electrode locations. The EABR thresholds in CNCs children were higher than those of children with normal cochlear nerve canals at electrode 11 and 22.

Wenjin W, Xiangrong T, Yun L, Jingrong L, Jianyong C, Xueling W, Zhiwu H, Hao W.
Neonatal hearing screening in remote areas of China: a comparison between rural and urban populations.
Objectives Universal neonatal hearing screening (UNHS) started late in some underdeveloped areas in China, with relatively scarce screening resources and a wide regional distribution. This study aimed to compare the screening performance between rural and urban populations, and to examine the characteristics and problems of UNHS in underdeveloped regions in China.
Methods A two-step hearing screening program was used in neonates born in Liuzhou Maternal and Child Health Hospital and in patients who were born in other hospitals, but admitted to the neonatal intensive care unit. This program involved distortion product otoacoustic emission and automated auditory brainstem response. Characteristics of each newborn, as well as the screening outcomes and performance were compared between rural and urban populations.

Results A total of 19,098 newborns were screened with a referral rate of 17.9% at the first step. Sixty-three (0.33%) newborns had hearing loss. The prevalence of permanent hearing loss was 2.25%. The average screening age was significantly older in the rural population than in the urban population in the first (P < 0.01) and second steps of screening (P < 0.05). The rural population had a higher referral rate in both steps than the urban population (P < 0.01). The follow-up rate was much lower in the rural population than in the urban population (P < 0.05), but dramatically increased in 2014 compared with the previous 2 years.

Conclusions A low follow-up rate is a critical issue when carrying out UNHS in developing countries, such as China, especially for rural populations. The government should establish more hearing referral centres to increase service coverage and supply financial assistance for low-income populations.


OBJECTIVE: The aim of this study was to analyze infants diagnosed with sensorineural or conductive hearing deficit and to identify risk factors associated with these defects.

MATERIAL AND METHODS: A retrospective analysis of infants diagnosed with hearing deficit based on the database of the universal newborn hearing screening program and medical records of the patients.

RESULTS: 27,935 infants were covered by the universal neonatal hearing screening program. 109 (0.39%) were diagnosed with hearing deficit and referred for treatment and rehabilitation. 56 (51.4%) children were diagnosed with conductive, 38 (34.9%) with sensorineural and 15 (13.8%) with mixed type of hearing deficit. Children with sensorineural hearing deficit more frequently suffered from hyperbilirubinemia (p < 0.05), while infants with conductive hearing loss were more frequently diagnosed with isolated craniofacial anomalies (p < 0.05). The prevalence of other risk factors did not differ between the groups. Sensorineural hearing deficit occurred almost 3 times more often bilaterally than unilaterally (p < 0.05). In other types of hearing deficit, the difference was not significant. In children with conductive and mixed type of hearing loss the impairment was mainly mild while among those with sensorineural hearing deficit in almost 45% it was severe and profound (p < 0.05). When analyzing the consistency between hearing screening test by means of otoacoustic emissions and the final diagnosis of hearing deficit we found that the highest agreement rate was observed in children with sensorineural hearing loss (p < 0.01).

CONCLUSIONS: The prevalence of most risk factors of hearing deficit was similar in children with sensorineural, conductive and mixed type of hearing loss, only hyperbilirubinemia seemed to predispose to sensorineural hearing deficit and isolated craniofacial malformations seemed to be associated with conductive hearing loss. Sensorineural hearing deficit usually occurred bilaterally and was severe or profound, while conductive and mixed type of hearing deficit were most often of mild degree. Most children with the final diagnosis of sensorineural hearing deficit had positive result of hearing screening by means of otoacoustic emissions.


OBJECTIVES: An effect of audio-visual (AV) integration is observed when the auditory and visual stimuli are incongruent (the McGurk effect). In general, AV integration is helpful especially in subjects wearing hearing aids or cochlear implants (CIs). However, the influence of AV integration on spoken word recognition in individuals with bilateral CIs (Bi-CIs) has not been fully investigated so far. In this study, we investigated AV integration in children with Bi-CIs.

METHODS: The study sample included thirty one prelingually deafened children who underwent sequential bilateral cochlear implantation. We assessed their responses to congruent and incongruent AV stimuli with three CI-listening modes: only the 1st CI, only the 2nd CI, and Bi-CIs. The responses were assessed in the whole group as well as in two sub-groups: a proficient group (syllable intelligibility ≥80% with the 1st CI) and a non-proficient group (syllable intelligibility < 80% with the 1st CI).

RESULTS: We found evidence of the McGurk effect in each of the three CI-listening modes. AV integration responses were observed in a subset of incongruent AV stimuli, and the patterns observed with the 1st CI and with Bi-CIs were similar. In the proficient group, the responses with the 2nd CI were not significantly different from those with the 1st CI whereas in the non-proficient group the responses with the 2nd CI were driven by visual stimuli more than those with the 1st CI.

CONCLUSION: Our results suggested that prelingually deafened Japanese children who underwent sequential bilateral cochlear implantation exhibit AV integration abilities, both in monaural listening as well as in binaural listening. We also observed a higher influence of visual stimuli on speech perception with the 2nd CI in the non-proficient group, sug-
suggesting that Bi-CIs listeners with poorer speech recognition rely on visual information more compared to the proficient subjects to compensate for poorer auditory input. Nevertheless, poorer quality auditory input with the 2nd CI did not interfere with AV integration with binaural listening (with Bi-CIs). Overall, the findings of this study might be used to inform future research to identify the best strategies for speech training using AV integration effectively in prelingually deafened children.

The report of the Polish Universal Neonatal Hearing Screening Program in 2016.
The Polish Universal Neonatal Hearing Screening Program (PUNHSP) has been carried out in Poland for 14 years. The main aim of this Program is to organize hearing screening tests and to gather the information about risk factors of hearing loss in almost all newborns in Poland. It consists of 496 centers at 3 referral levels. A total of 5 458 114 children had been registered in the Central Database (CDB) of PUNHSP by the 22nd of August 2017. Bilateral sensorineural hearing loss was the most frequently appearing hearing impairment in children. It was diagnosed in 260 cases in 2016. This report presents the most important results and conclusions concerning the running of the PUNHSP in 2016.