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- To bring together a range of recently-published research reports, articles and electronic resources to help all staff keep up-to-date with research and practice.

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35. Genotyping and audiological characteristics of infants with a single-allele SLC26A4 mutation.

36. Otosyphilis: Resurgence of an Old Disease.


38. Auditory Evoked Potential Inconsistency in Sudden Unilateral Hearing Loss with Multiple Sclerosis.

39. The evaluation of the sense of hearing in patients with carotid artery stenosis within the extracranial segments.

40. Congenital cytomegalovirus infection inducing non-congenital sensorineural hearing loss during childhood; a systematic review.

41. Predicting sequential bilateral cochlear implantation performance in postlingually deafened adults; A retrospective cohort study.

42. Sound therapy (using amplification devices and/or sound generators) for tinnitus.

Author(s): Dupuis, Kate; Yusupov, Iris; Vandezmorris, Susan; Murphy, Kelly L; Rewilak, Dmytro; Stokes, Kathryn A; Reed, Marilyn

Source: Canadian journal on aging = La revue canadienne du vieillissement; Jun 2019; vol. 38 (no. 2); p. 245-252

Publication Date: Jun 2019

Publication Type(s): Journal Article

PubMedID: 30522534

Abstract:

Hearing loss is highly prevalent in older adults and can pose challenges for neuropsychologists, as assessment and intervention procedures often involve orally presented information which must be accurately heard. This project examined the hearing status of 20 clients (mean age = 71 years) in a hospital-based outpatient neuropsychology clinic, and explored whether information about hearing loss informed neuropsychologists’ clinical practice. A research assistant administered a brief hearing screening test to each participant. Four treating neuropsychologists were asked to comment on their client’s hearing status before and after being shown their client’s hearing screen test results. Screening revealed that the majority of participants had at least mild hearing loss, and that the neuropsychologists were relatively accurate (60%) at estimating their clients’ hearing status. Neuropsychologists used information about a client’s hearing status to make recommendations that clients pursue audiologic services, and to educate clients and family members about hearing loss and communication.

Database: Medline

2. Frequency of auditory involvement and of associated factors in patients with juvenile idiopathic arthritis.

Author(s): Céspedes Cruz, Adriana Ivonne; Méndez Núñez, Myriam; Solís Vallejo, Eunice; Zeferino Cruz, Maritza; Torres Jiménez, Alfonso Ragnar; Ocampo Sánchez, Verónica; Flores Meza, Beatriz; Quintana Ruiz, Norma

Source: Reumatologia clinica; 2019; vol. 15 (no. 3); p. 152-155

Publication Date: 2019

Publication Type(s): Journal Article

PubMedID: 28923429

Abstract:

INTRODUCTION: Juvenile idiopathic arthritis (JIA) is a chronic autoimmune disease characterized by the presence of arthritis in children under 16 years of age for more than 6 weeks in the absence of any other known cause. The extra-articular manifestations, especially in the audiovestibular system, are related to the involvement of the joints of the ossicular chain as a result of the inflammatory process in the synovium. Previous clinical studies in pediatric patients have shown conductive or sensorineural hearing loss.
OBJECTIVE: The aim of this study was to assess the frequency of hearing impairment and of associated factors in patients with JIA.

METHODOLOGY: A prospective, analytical study was conducted from January 2013 to August 2014 in 62 patients with JIA aged between 5 and 15 years. The study was approved by the local ethics committee and parents signed their informed consent. All subjects underwent audiological examination involving otomicroscopy, audiometry, tympanometry, stapedius reflex and test for transient otoacoustic emissions (TOAE); rheumatologic evaluation included joint examination and the application of a measure of functional ability (disability) using the Childhood Health Assessment Questionnaire (CHAQ). Measures of central tendency and of dispersion were used (chi-square for associations and P<.05 for statistical significance).

RESULTS: Sixty-two patients were included: 56 girls and 6 boys, mean age 11.9 years and mean disease duration of 3.4 years; 46% had rheumatoid factor (RF)-positive polyarticular JIA, 40% had RF-negative polyarticular JIA, 15% had disease of systemic onset and 3% had oligoarthritis. Active disease was found in 29 patients and 33 were in remission with medication. Of the total of 124 ears evaluated according to the Jerger classification for tympanometry, abnormal findings were observed in 78 that were type As and in 1 that was type Ad, whereas there were 45 type A ears. Hearing loss was disclosed by speech audiometry, rather than by pure tone audiometry. The TOAE were absent in 4% of those assessed and the stapedius reflex was absent in less than 10%. Factors that had a positive correlation with hearing impairment were RF-positive polyarticular JIA, disease duration, degree of disability and the erythrocyte sedimentation rate level (P<.000).

CONCLUSION: The presence of an abnormal tympanogram suggested early involvement in the structure of the tympanic-ossicular complex; however, 3.4 years later, no hearing loss had been reported.

Database: Medline

3. Diagnostic accuracy of non-specialist versus specialist health workers in diagnosing hearing loss and ear disease in Malawi.

Author(s): Bright, Tess; Mulwafu, W; Phiri, M; Ensink, R J H; Smith, A; Yip, J; Mactaggart, I; Polack, S

Source: Tropical medicine & international health : TM & IH; Apr 2019

Publication Date: Apr 2019

Publication Type(s): Journal Article

PubMedID: 31001894

Available at Tropical medicine & international health : TM & IH - from Wiley Online Library

Abstract:

OBJECTIVE: To determine whether a non-specialist health worker can accurately undertake audiometry and otoscopy, the essential clinical examinations in a survey of hearing loss, instead of a highly skilled specialist (i.e. ENT or audiologist).

METHODS: Clinic-based diagnostic accuracy study in Malawi. Consecutively sampled participants ≥18 years had their hearing tested using a validated tablet-based audiometer (hearTest) by an audiologist (gold-standard), an audiology officer, a nurse and a community health worker (CHW). Otoscopy for diagnosis of ear pathologies was conducted by an ENT specialist (gold-standard), an ENT clinical officer, a CHW, an ENT nurse, and a general nurse. Sensitivity, specificity and kappa (k) were calculated. 80% sensitivity, 70% specificity, and kappa of 0.6 were considered adequate.
RESULTS: 617 participants were included. High sensitivity (>90%) and specificity (>85%) in detecting bilateral hearing loss was obtained by all non-specialists. For otoscopy, sensitivity and specificity were >80% for all non-specialists in diagnosing any pathology except for the ENT nurse. Agreement in diagnoses for the ENT clinical officer was good (k=0.7) in both ears. For other assessors, moderate agreement was found (k=0.5).

CONCLUSION: A non-specialist can be trained to accurately assess hearing using mobile-based audiometry. However, accurate diagnosis of ear conditions requires at least an ENT clinical officer (or equivalent). Conducting surveys of hearing loss with non-specialists could lower costs and increase data collection, particularly in low and middle-income countries, where ENT specialists are scarce. This article is protected by copyright. All rights reserved.

Database: Medline

4. Is one of these two techniques: CO2 laser versus microdrill assisted stapedotomy results in better post-operative hearing outcome?

Author(s): Altamami, Nasser M; Huyghues des Etages, Gunther; Fieux, Maxime; Coudert, Aurélie; Hermann, Ruben; Zaouche, Sandra; Truy, Eric; Tringali, Stéphane

Source: European archives of oto-rhino-laryngology : official journal of the European Federation of Oto-Rhino-Laryngological Societies (EUFOS) : affiliated with the German Society for Oto-Rhino-Laryngology - Head and Neck Surgery; Apr 2019

Publication Date: Apr 2019
Publication Type(s): Journal Article
PubMedID: 30949824

Abstract:

OBJECTIVE: To evaluate hearing results and outcome using two different surgical techniques (microdrill and CO2 Laser fenestration) in the treatment of conductive hearing loss in patients with otosclerosis.

STUDY DESIGN: Retrospective audiometric database and chart review from January 2005 until December 2016.

SETTING: Two tertiary referral hospitals

MATERIALS AND METHODS: Seven-hundred forty-two primary stapedotomy have been reviewed retrospectively in two referral hospitals. This multicenter study compared 424 patients operated for otosclerosis with microdrill technique and 318 patients operated with CO2 laser assisted stapedotomy. Preoperative and postoperative audiological assessment (following the recommendations of the Committee on Hearing and Equilibrium) were compared between the two groups at least 6 weeks and at 1 year or more. Measure of overclosure and hearing damage have been analyzed and compared between the groups.

RESULTS: There were no statistically significant differences in demographic data between the two groups and no statistically significant difference in hearing outcome between the two groups. CO2 Laser with 0.4 piston showed slightly better results to close the air-bone gap postoperatively to ≤10 dB (84% as compared with the 80% of patients operated with microdrill technique). Patients operated with microdrill technique and 0.6 piston have less damage to hearing at 4 kHz.

CONCLUSION: The use of CO2 laser seems associated with better postoperative air-bone gap closure. However, it carries more risk of hearing damage at 4 kHz at it is the case for the microdrill at 1 kHz. In general, postoperative hearing outcome using these two surgical techniques is comparable.

**Author(s):** Neumann, Katrin; Thomas, Jan Peter; Voelter, Christiane; Dazert, Stefan

**Source:** International journal of pediatric otolaryngology; Apr 2019; vol. 122; p. 117-125

**Abstract:**

**OBJECTIVES:** Bone conduction hearing devices integrated in softbands (BCDSs) are frequently not well accepted by children with conductive hearing loss due to pressure on the head, sweating, or cosmetic stigma. A non-surgical hearing system (ADHEAR) uses a new bone conduction concept consisting of an audio processor connected to an adhesive adapter fixed behind the ear. This study is the first to evaluate the audiological and clinical outcome of this novel system, comparing it with conventional BCDSs in a short- and mid-term follow-up in children under 10 years of age.

**METHODS:** The ADHEAR was compared to a BCDS in 10 children with conductive hearing loss (age: 0.7-9.7 years). Aided and unaided pure tone/behavioral observational audiometry and, if applicable, speech audiometry in quiet and noise were performed initially with both devices and after 8 weeks with the ADHEAR alone. The subjective hearing gain and usage of the new hearing system, as well as patients' and parents' satisfaction were assessed using questionnaires.

**RESULTS:** The functional gain with the ADHEAR averaged over 0.5, 1, 2, and 4 kHz exceeded that of the conventional BCDS (35.6 dB ± 15.1 vs. 29.9 dB ± 14.6, p = .001, n = 9 ears). Speech perception in quiet and noise (n = 8) improved in the aided situation similarly for both hearing devices. The parents of 8 of 10 children evaluated the ADHEAR system as being useful. Minor wearing problems occurred occasionally. Eight children continued using the ADHEAR after the study, one received an active middle ear implant and one continued to use a BCDS.

**CONCLUSION:** The ADHEAR system is a promising solution for children with conductive hearing loss or chronically draining ears.

**Database:** Medline

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6. Avoiding Furosemide Ototoxicity Associated With Single-Ventricle Repair in Young Infants.

**Author(s):** Robertson, Charlene M T; Bork, Karin T; Tawfik, Gerda; Bond, Gwen Y; Hendson, Leonora; Dinu, Irina A; Khodayari Moez, Elham; Rebeya, Ivan M; Garcia Guerra, Gonzalo; Joffe, Ari R

**Source:** Pediatric critical care medicine : a journal of the Society of Critical Care Medicine and the World Federation of Pediatric Intensive and Critical Care Societies; Apr 2019; vol. 20 (no. 4); p. 350-356

**Publication Date:** Apr 2019

**Abstract:**
OBJECTIVE: To reduce bilateral delayed-onset progressive sensory permanent hearing loss using a systems-wide quality improvement project with adherence to best practice for the administration of furosemide.

DESIGN: Prospective cohort study with regular audiologic follow-up assessment of survivors both before and after a 2007-2008 quality improvement practice change.

SETTING: The referral center in Western Canada for complex cardiac surgery, with comprehensive multidisciplinary follow-up by the Complex Pediatric Therapies Follow-up Program.

PATIENTS: All consecutive patients having single-ventricle palliative cardiac surgery at age 6 weeks old or younger.

INTERVENTIONS: A 2007-2008 quality improvement practice change consisted of a Parenteral Drug Monograph revision indicating slow IV administration of furosemide, an educational program, and an evaluation.

MEASUREMENTS AND MAIN RESULTS: The outcome measure was the prevalence of permanent hearing loss by 4 years old. Firth multiple logistic regression compared pre (1996-2008) to post (2008-2012) practice change occurrence of permanent hearing loss, adjusting for confounding variables, including all hospital days, extracorporeal membrane oxygenation, cardiopulmonary bypass time, age at first surgery, dialysis, and sepsis. From 1996 to 2012, 259 infants had single-ventricle palliative surgery at age 6 weeks old or younger, with 173 (64%) surviving to age 4 years. Of survivors, 106 (61%) were male, age at surgery was 11.6 days (9.0 d), and total hospitalization days by age 4 years were 64 (42); 18 (10%) had cardiopulmonary resuscitation and 38 (22%) had sepsis at any time. All 173 (100%) had 4-year follow-up. Pre- to postpractice change permanent hearing loss dropped from 17/100 (17%) to 0/73 (0%) of survivors. On Firth multiple logistic regression, the only variable statistically associated with permanent hearing loss was the pre- to postpractice change time period (odds ratio, 0.03; 95% CI, 0-0.35; p = 0.001).

CONCLUSIONS: A practice change to ensure slow IV administration of furosemide eliminated permanent hearing loss. Centers caring for critically ill infants, particularly those with single-ventricle anatomy or hypoxia, should review their drug administration guidelines and adhere to best practice for administration of IV furosemide.

Database: Medline

7. How Do We Know That Our Patients Have Benefitted From Our ENT/Audiological Interventions?
Presented at the Annual Meeting of ADANO 2016 in Berlin.

Author(s): Hall, Deborah A; Kitterick, Pádraig; Heffernan, Eithne; Fackrell, Kathryn; Lucas, Laura; Ferguson, Melanie

Source: Otology & neurotology : official publication of the American Otological Society, American Neurotology Society [and] European Academy of Otology and Neurotology; Apr 2019; vol. 40 (no. 4); p. e474

Publication Date: Apr 2019

Publication Type(s): Journal Article

PubMedID: 30870383

Abstract: This short review article gives an introduction to some of the fundamental concepts and challenges facing measurement in hearing healthcare practice and research. The impact of hearing loss almost always extends beyond the sensory impairment itself, even when the measured degree of audiometric loss is mild. Yet, going beyond audibility, into the realm of measuring impact, takes us into a much more complex and less well-defined space. How does one therefore best measure the
therapeutic benefit for evaluating efficacy or for clinical practice audit? Three case studies illustrate approaches to overcome such challenges. Each example highlights the importance of thinking critically about what it is one is seeking to try to measure, rather than selecting a questionnaire instrument based simply on its popularity or accessibility. We conclude by highlighting the important role that clinicians can play in collecting clinical data about their preferred instruments so that we have some evidence to inform decisions about good practice (content validity etc.). We would also strongly support open data sharing as we think that this is one of the best ways to make the most rapid progress the field.

Database: Medline

8. Bilateral duplication of the internal auditory canals and bilateral cochlear implant outcomes and review.

Author(s): Thompson, Murray R; Birman, Catherine S

Source: International journal of pediatric otorhinolaryngology; Apr 2019; vol. 119; p. 41-46

Publication Date: Apr 2019

Publication Type(s): Case Reports Journal Article Review

PubMedID: 30665175

Abstract:

OBJECTIVES: Bilateral duplication of the internal auditory canal (IAC) is rare and is associated with profound sensorineural hearing loss. The present study aims to review our experience with bilateral cochlear implantation (CI) in children with a duplication of the IAC and to review the literature.

METHODS: The Sydney Cochlear Implant Centre database was searched for children with duplication of the internal auditory canal. Data was collected regarding clinical history, MRI and CT findings, auditory brainstem responses (ABR), tympanometry and otoacoustic emissions (OAE), visually reinforced orientation audiometry, auditory brainstem response, electrocochleography (ECochG), transtympanic electrical auditory brainstem response (ABR), aided cortical evoked potentials (CAEP) and intraoperative neural response telemetry (NRT) and CI evoked electrical auditory brainstem testing.

RESULTS: two children with bilateral duplication of the IAC were identified who successfully underwent bilateral cochlear implantation. Audiological development was monitored for 2 and 3 years respectively, both children could spontaneously verbalise and displayed Categories of Auditory Performance (CAP) score of 5 and 6 respectively.

CONCLUSION: Children with duplication of the IAC, with accompanying cochlear nerve dysplasia (CND) can benefit from CI surgery, and verbal receptive and expressive language is possible.

Database: Medline


Author(s): Ziąbka, Magdalena; Malec, Katarzyna

Source: Expert review of medical devices; Apr 2019; vol. 16 (no. 4); p. 325-331

Publication Date: Apr 2019

Publication Type(s): Journal Article
Abstract:

OBJECTIVE: Otitis media is one of the most common illnesses which may cause ossicles destruction and conductive hearing loss. However, nowadays the damaged middle ear bones may be replaced by a partial or total ossicular replacement prosthesis. The main aim of this article was to confirm the proper functioning of the new middle ear prosthesis.

METHODS: This work describes first clinical trials conducted on a group of three patients with the case of interrupted ossicles continuity and chronic otitis media. The clinical trials were performed according to permission No. 157/KBL/OIL2016. The patients were subjected to the bones chain reconstruction via implanting a bactericidal middle ear prosthesis called Otoimplant. The following preoperative and postoperative parameters have been evaluated: Air-Bone-Gap values, mean ABG values in different frequencies, bone and air conductivity, speech audiometry and microbiological assessment. The patients' recovery was observed according to the study protocol on the 7th day and 1, 3, 6 and 12 months after the surgery.

RESULTS: Audiological tests revealed that the mean Air-Bone-Gap was reduced by 36% in all the patients after 1 year. No bacteria or fungi were found in the middle ear spaces.

CONCLUSIONS: Results confirm the bactericidal efficacy and hearing improvement of the Otoimplant.

Database: Medline


Author(s): Tovi, H; Ovadia, H; Eliashar, R; de Jong, M A; Gross, M

Source: European annals of otorhinolaryngology, head and neck diseases; Apr 2019; vol. 136 (no. 2); p. 99-101

Publication Date: Apr 2019

Publication Type(s): Journal Article

PubMedID: 30606654

Abstract:

OBJECTIVES: To define the clinical association of serum prestin autoantibodies and their impact on prognosis, as specific serum diagnostic markers in patients with idiopathic sudden sensorineural hearing loss (ISSNHL).

DESIGN: Sera from 63 patients with ISSNHL were screened prospectively for the presence of prestin autoantibodies by an enzyme-linked immunosorbent assay (Elisa) test. Serum was assayed for anti-prestin IgG antibodies using recombinant human prestin (SLC26 A5). Demographic, clinical, and audiometric variables were analyzed.

RESULTS: Two patients (3.17%) had demonstrable anti-prestin antibodies in serum (exact 95% CI: 1.16% to 7.5%). No statistically significant association was found between prestin autoantibodies and demographic or audiologic parameters.

CONCLUSIONS: This preliminary and novel study does not support the presence of an active humoral immune reaction against prestin in ISSNHL.

Database: Medline
11. Results of a Targeted Screening Program for Congenital Cytomegalovirus Infection in Infants Who Fail Newborn Hearing Screening.

**Author(s):** Vancor, Emily; Shapiro, Eugene D; Loyal, Jaspreet  
**Source:** Journal of the Pediatric Infectious Diseases Society; Mar 2019; vol. 8 (no. 1); p. 55-59  
**Publication Date:** Mar 2019  
**Publication Type(s):** Journal Article  
**PubMedID:** 29373759  
**Abstract:**
**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.

**Database:** Medline

12. Integration of congenital cytomegalovirus screening within a newborn hearing screening programme.

**Author(s):** Beswick, Rachael; David, Michael; Higashi, Hideki; Thomas, Delene; Nourse, Clare; Koh, Guan; Koorts, Pieter; Jardine, Luke A; Clark, Julia E  
**Source:** Journal of paediatrics and child health; Mar 2019  
**Publication Date:** Mar 2019  
**Publication Type(s):** Journal Article  
**PubMedID:** 30916438  
Available at [Journal of paediatrics and child health](https://onlinelibrary.wiley.com/doi/abs/10.1111/jpc.13466) - from Wiley Online Library  
**Abstract:**
**AIM:** Targeted screening by a salivary cytomegalovirus (CMV) polymerase chain reaction (PCR) of infants who ‘refer’ on their newborn hearing screen has been suggested as an easy, reliable and cost-effective approach to identify and treat babies with congenital CMV (cCMV) to improve hearing
outcomes. This study aimed to investigate the feasibility and cost-effectiveness of introducing targeted salivary cCMV testing into a newborn hearing screening programme.

**METHODS:** The study included three tertiary maternity hospitals in Queensland, Australia between August 2014 and April 2016. Infants who 'referred' on the newborn hearing screen were offered a salivary swab for CMV PCR at the point of referral to audiology. Swabs were routinely processed and tested for CMV DNA by real-time quantitative PCR. Parents of babies with a positive CMV PCR were notified, and the babies were medically assessed and, where appropriate, were offered treatment (oral valganciclovir).

**RESULTS:** Of eligible infants, the parents of 83.0% (234/283) consented to the cCMV screen. Of these, 96.6% returned a negative result (226/234), and 3.4% (8/234) returned a positive result (three true positive; five false positive). The prevalence of cCMV for infants with confirmed hearing loss was 3.64% (P = 2/55; confidence interval = 0.44-12.53%). The cost comparison suggests the cost implementation of cCMV screening (and subsequent potential treatment benefits and management over time), compared to non-screening (and subsequent management), to be negligible.

**CONCLUSION:** Incorporating cCMV testing into Universal Newborn Hearing Screening within Queensland is realistic and achievable, both practically and financially.

**Database:** Medline

13. A novel pathogenic variant in OSBPL2 linked to hereditary late-onset deafness in a Mongolian family.

**Author(s):** Wu, Ningjin; Husile, Husile; Yang, Liqing; Cao, Yaning; Li, Xing; Huo, Wenyan; Bai, Haihua; Liu, Yangjian; Wu, Qizhu

**Source:** BMC medical genetics; Mar 2019; vol. 20 (no. 1); p. 43

**Publication Date:** Mar 2019

**Publication Type(s):** Journal Article

**PubMedID:** 30894143

Available at [BMC medical genetics](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6402698/) - from BioMed Central

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Available at [BMC medical genetics](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6402698/) - from ProQuest (Health Research Premium) - NHS Version

Available at [BMC medical genetics](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6402698/) - from Unpaywall

**Abstract:**

**BACKGROUND:** To investigate the clinical features and the underlying causal gene of a family with hereditary late-onset deafness in Inner Mongolia of China, and to provide evidence for the early genetic screening and diagnosis of this disease.

**METHODS:** Family data were collected to draw a pedigree. Audiological testing and physical examination of the family members were conducted following questionnaire. Genomic DNA was extracted from peripheral blood of 5 family members (3 patients and 2 normal control) and subjected to whole genome sequencing for identifying deafness casual genes. The pathogenic variant in the deafness gene was further confirmed by Sanger sequencing.

**RESULTS:** The family is composed of a total of 6 generations, with 53 traceable individuals. In this family, 19 of them were diagnosed with post lingual deafness with the age of onset between 10 and
40 years, displaying delayed and progressive hearing loss. Patients with hearing loss showed bilateral symmetry and mild to severe sensorineural deafness. The pattern of deafness inheritance in this family is autosomal dominant. Whole genome sequencing identified a novel pathogenic frameshift mutation, c.158_159delAA (p.Gln53Arg fs*100) in the gene OSBPL2 (Oxysterol-binding protein-related protein 2, NM_144498.2), which is absent from genomic data of 201 unrelated normal subjects. This pathogenic variant was further validated by Sanger sequencing, and was found to cosegregate in this family.

CONCLUSIONS: Whole genome sequencing identified a two-nucleotide deletion in OSBPL2 (c.158_159delAA) as the pathogenic variant for deafness in the family. Our finding expands the mutational spectrum of OSBPL2 and contributes to the pathogenic variant list in genetic counseling for deafness screening.

Database: Medline


Author(s): Alkahtani, Rania; Rowan, Daniel; Kattan, Nad; Alwan, Nisreen A
Source: International journal of pediatric otorhinolaryngology; Mar 2019; vol. 122; p. 27-34
Publication Date: Mar 2019
Publication Type(s): Journal Article
PubMedID: 30933841
Abstract:
OBJECTIVES: To identify the average age of identification (AOI) and characteristics of Saudi children with sensorineural hearing loss (SNHL).
METHODS: Two cross-sectional studies were undertaken. Study A: the medical records of 1166 children aged 0-10 years old who visited the audiology clinics in four hospitals in Riyadh and Dammam during 2015 were reviewed. Study B: 174 carers of children aged 0-12 years who visited the audiology clinics in four hospitals in Riyadh during a three-month period were surveyed.
RESULTS: The mean AOI with SNHL in children was 3.2 years (SD = 2.5 years) and 3.1 years (SD = 2.6 years) with 14% and 16% not identified until after primary school age for Studies A and B, respectively. The presence of SNHL was positively associated with parental consanguinity, positive family history of SNHL, history of chemotherapy treatment, brain pathology and prior parental concern regarding their child’s hearing.
CONCLUSION: AOI of SNHL among Saudi children is deemed high in relation to the likely age of onset, with about 15 in 100 children identified after school age. Childhood hearing screening programmes (at birth and at school entry) should be considered in order to intervene earlier.
Database: Medline


Author(s): Iwanicka-Proniccka, Katarzyna; Ciara, Elżbieta; Piekutowska-Abramczuk, Dorota; Halat, Paulina; Pajdowska, Magdalena; Pronicki, Maciej
Source: International journal of pediatric otolaryngology; Mar 2019; vol. 121; p. 143-149
OBJECTIVES: Although hearing loss is a well-known symptom of mitochondria-related disorders, it is not clear how often it is a congenital and cochlear impairment. The Newborn Hearing Screening Program (NHSP) enables to distinguish congenital cochlear deafness from an acquired hearing deficit. The initial aim of the study was to research the frequency of the congenital cochlear hearing loss among patients with various gene defects resulting in mitochondrial disorders. The research process brought on an additional gain: basing on our preliminary study group of 80 patients, in 12 patients altogether we identified two defected genes responsible for mitochondrial disorders, whose carriers did not pass the NHSP. Finally, these patients were diagnosed with the congenital cochlear deafness.

MATERIAL AND METHODS: The results of the NHSP in the patients with mitochondrial disorders diagnosed in our tertiary reference center were analyzed. Only the cases with confirmed mutations were qualified for the study group. The NHSP database included 80 patients with mutations in 31 different genes: 25 nuclear-encoded and 6 mtDNA-encoded. We searched the literature for the presence of a congenital hearing impairment (CHI) in mitochondrial disorders caused by changes in 278 already known genes.

RESULTS: For 68 patients from the study group the NHSP test indicated a proper cochlear function and thus suggested normal hearing. For 12 mitochondrial patients, the NHSP test indicated the requirement for the further audiological diagnosis, and finally CHI was confirmed in 8 of them. This latter subset included patients with pathogenic variants in RRM2B and SERAC1, known as "deafness-causing genes". Contrary to our initial expectations, the patients carrying mutations in other "deafness-causing genes": MPV17, POLG, COX10, as well as other mitochondria-related genes, all reported in literature, did not indicate any CHI following the NHSP test.

CONCLUSION: Our study indicates that the cochlear CHI is a phenotypic feature of the RRM2B and SERAC1 related defects. The diagnosis of the CHI following the NHSP allows to early distinguish those defects from other mitochondria-related disorders in which the NHSP test result is correct. Wider studies are needed to assess the significance of this observation.

Database: Medline

16. The atherogenic index (ATH index) as a potential predictive marker of idiopathic sudden sensorineural hearing loss: a case control study.

Author(s): Kaneva, Anastasiya M; Yanov, Yury K; Bojko, Svetlana G; Kudryavykh, Olga E; Potolitsyna, Natalya N; Bojko, Evgeny R; Odland, Jon Ø

Source: Lipids in health and disease; Mar 2019; vol. 18 (no. 1); p. 64

Publication Date: Mar 2019
Publication Type(s): Journal Article
PubMedID: 30876416
Available at Lipids in health and disease - from BioMed Central
Available at Lipids in health and disease - from Europe PubMed Central - Open Access
Available at Lipids in health and disease - from EBSCO (MEDLINE Complete)
BACKGROUND: The importance of blood lipids in the pathogenesis of sudden sensorineural hearing loss (SSNHL) is widely discussed in the literature. However, the published results that hyperlipidaemia causes hearing problems are contradictory. The objective of this study was to establish whether increased lipid levels affect the risk of idiopathic SSNHL.

METHODS: A case-controlled study was conducted of 27 patients with idiopathic SSNHL and 24 healthy control subjects. All of the subjects underwent complete audiological examination. The plasma levels of total cholesterol (TC), triglycerides (TG), high-density lipoprotein cholesterol (HDL-C), apolipoprotein (apo) A-I, apoB and apoE were measured with commercially available kits (Chronolab Systems, Spain). Several clinical ratios and indices of lipid metabolism were calculated.

RESULTS: Detailed analysis of lipid metabolism in patients with idiopathic SSNHL has shown that disturbances in auditory function are associated with increased atherogenicity of the lipid profile. However, there were no significant differences in the conventional parameters of lipid metabolism (TC, TG and HDL-C) between patients with idiopathic SSNHL and subjects in the control group. Higher values of the apoB/apoA-I ratio, atherogenic index of plasma (AIP) and atherogenic index (ATH index) in patients with SSNHL indicated increased atherogenicity of the lipid profile. Binary logistic regression analysis showed that of these three indices, only higher values of the ATH index were significantly associated with an increased risk of idiopathic SSNHL.

CONCLUSIONS: The ATH index can be used as a marker indicating the risk of idiopathic SSNHL when the conventional lipid indices are still normal.

Database: Medline
Design: The design of this study is a single-site, prospective, double-blind clinical trial. Outcome measures were obtained after a typical 4- to 5-week trial period. An optional follow-up of a 4-week audiology-based (AB) best practices trial was also included for replication and comparison purposes. Setting Older adults from the general community were recruited via newspaper and community flyers to participate at a university research clinic.

Participants: Participants were adults, aged 54-78 years, with mild-moderate hearing loss. Forty-one participants enrolled as a volunteer sample; 40 completed the intervention. Intervention(s) All participants received the same high-end digital mini-behind-the-ear hearing aids fitted bilaterally. CD participants self-selected their own preprogrammed hearing aids via an OTC-type model. One of the 3 devices from which participants could choose was programmed to be a placebo device with no functional gain.

Primary and Secondary Outcome Measures: The primary outcome measure is the 66-item self-report Profile of Hearing Aid Benefit (Cox & Alexander, 1990). The secondary outcome measure is the Connected Speech Test (Cox, Alexander, & Gilmore, 1987) benefit. Additional measures of hearing aid benefit and usage were also obtained.

Results: Per-protocol analyses based on the data from the 40 (of 41) participants who completed the study were performed. Hearing aid outcomes from this follow-up CD (CD2) cohort were positive and generally the same as for the original CD cohort. CD service delivery model was efficacious relative to CD-based placebo control, with medium effect sizes observed. Approximately half of the CD2 group was likely to purchase hearing aids after the trial, similar to findings for the original CD cohort. Outcomes improved significantly for the 32 CD2 participants who elected to complete the optional 4-week AB trial. For this largely unscreened sample, more individuals with healthy hearing sought amplification, and many of these individuals (35%) chose placebo devices for both ears.

Conclusions: Prior positive outcomes for CD service delivery have been replicated in a less restrictive approach to participant recruitment. The CD approach was again found to be efficacious. Significantly better outcomes were observed after subsequent AB service delivery follow-up, also replicating prior findings. Efficacious OTC models, including those using similar CD approaches to hearing aid self-selection, may increase accessibility and affordability of hearing aids for millions of older adults. Front-end guidance to consumers regarding the best path to intervention, ranging from self-screening of hearing online to a full audioligic assessment, appears to be critical to optimize the success of OTC approaches. Trial Registration: Clinicaltrials.gov: NCT01788432; https://clinicaltrials.gov/ct2/show/NCT01788423 Supplemental Material https://doi.org/10.23641/asha.7728479.

Database: Medline

18. Audiological evaluation of infants using mother’s voice.

Author(s): Saito, Osamu; Nishimura, Tadashi; Morimoto, Chihiro; Otsuka, Shintaro; Uratani, Yuka; Matsunaga, Yasuko; Hosoi, Hiroshi; Kitahara, Tadashi

Source: International journal of pediatric otorhinolaryngology; Mar 2019; vol. 121; p. 81-87

Publication Date: Mar 2019

Publication Type(s): Journal Article

PubMedID: 30877980

Abstract:
OBJECTIVES: Hearing loss is a serious problem in infants and children because it may interfere with the development of typical speech, verbal language, and auditory and communication skills. By measuring hearing ability (thresholds) as early as possible, even as early as during infancy, effective treatment can be administered. These treatments may significantly reduce the handicap associated with hearing loss. However, at times during behavioral auditory tests, observers cannot determine whether or not an accurate threshold was obtained. To support the use of infant audiometry for accurate diagnosis, audiologic behavioral responses may be obtained by selecting stimuli that interest infants, e.g., their mothers' voices.

METHODS: We evaluated 30 infants who were presented to our hospital for behavioral auditory assessment in 2016. The infants' ages ranged from 4 months to 3 years and 6 months. The mean age was 1 year and 10 months ±10 months (±standard deviation). The infants' hearing thresholds for their mothers' voices and warble tones at 250-4000 Hz were measured. Auditory brainstem response (ABR) had already been evaluated in 24 infants. Relationships between mother’s voice and warble tone or ABR thresholds as well as responses to the initial stimuli and stimuli at the threshold were investigated. These responses were classified into four grades (no response, uncertain response, possible positive response, and positive response), and the response to mother’s voice and warble tone were subsequently compared.

RESULTS: Mother’s voice thresholds significantly correlated with all warble tone thresholds. In the relationship between the mother's voice threshold and average hearing levels of 500, 1000, and 2000 Hz, two infants were outliers. In these infants, the average hearing levels were relatively higher than the mother's voice thresholds. Judging from their ABR thresholds, the mother’s voice thresholds were valid and the average hearing levels were worse than their original assessed hearing ability. The responses to mothers' voices were more distinct than those to warble tones, both for initial stimuli presentation and the determined threshold.

CONCLUSIONS: Audiologic behavioral responses to mothers’ voices were clearer than those for warble tones. Evaluations that use the mother’s voice threshold are useful for estimating hearing levels in infants.

Database: Medline

19. [Caloric test and hearing characteristics in patients with vestibular migraine and Meniere's disease].

Author(s): Yu, X; Wang, L Y; Han, W; Tian, L J; Liu, X L

Source: Lin chuang er bi yan hou tou jing wai ke za zhi = Journal of clinical otorhinolaryngology, head, and neck surgery; Mar 2019; vol. 33 (no. 3); p. 228-231

Publication Date: Mar 2019

Publication Type(s): English Abstract Journal Article

PubMedID: 30813691

Abstract:

Objective: This study was designed to distinguish vestibular migraine (VM) from Meniere's disease (MD) by comparing age, sex, family history, audiological and vestibular test results in patients with VM or MD.

Method: Forty-six patients with suspected VM, 60 patients with confirmed VM, and 60 patients with confirmed MD were studied. All patients were asked for detailed medical history, and then
underwent bithermal caloric test and audiological test. The hearing function and vestibular function were analyzed.

**Result:** ① General conditions: the sex ratio with suspected VM, confirmed VM and confirmed MD was 1:4.75, 1:5.67 and 1:1. Family history in these three groups was 80.43% (37/46), 66.67% (40/60) and 6.67% (4/60), respectively. ② In the three groups, the abnormal rates of caloric test were 50.00% (23/46), 31.67% (19/60) and 78.33% (47/60), respectively. ③ The rate of pure tone audiometry abnormality in the three groups was 60.87% (28/46), 63.33% (38/60) and 100.00% (60/60), respectively. Of 46 suspected VM patients, 18 (39.13%) had high frequency hearing loss, 7 (15.22%) had full frequency hearing loss, and 3 (6.52%) had low frequency hearing loss. Among 60 confirmed VM patients, 18 (30.00%) had high frequency hearing loss, 15 (25.00%) had full frequency hearing loss, 4 (6.67%) had low frequency hearing loss, and 1 case (1.67%) had low frequency and high frequency hearing loss. Among 60 confirmed MD patients, 56 (93.33%) had full frequency hearing loss, and 4 (6.67%) had low frequency hearing loss. ④ The difference of audiological test between patients with confirmed VM and patients with suspected VM was not statistically significant (P>0.05). The difference of bithermal caloric test between patients with confirmed VM and patients with suspected VM was not statistically significant (P>0.05). Compared with MD patients, the incidence of hearing loss and the incidence of abnormal bithermal caloric test in VM patients is lower (P<0.05).

**Conclusion:** The abnormality rate of caloric test and the incidence of hearing loss in MD patients were higher than VM. The confirmed and suspected VM patients can be accompanied by hearing loss mainly with high hearing frequency decline. There was no difference in clinical characteristics between confirmed and suspected VM patients.

**Database:** Medline

### 20. Interregional Newborn Hearing Screening via Telehealth in Ghana.

**Author(s):** Ameyaw, Graham Amponsah; Ribera, John; Anim-Sampong, Samuel

**Source:** Journal of the American Academy of Audiology; Mar 2019; vol. 30 (no. 3); p. 178-186

**Publication Date:** Mar 2019

**Publication Type(s):** Journal Article

**PubMedID:** 30461394

**Abstract:**

**BACKGROUND:** Newborn hearing screening is a vital aspect of the Early Hearing Detection and Intervention program, aimed at detecting hearing loss in children for prompt treatment. In Ghana, this kind of pediatric hearing service is available at only one health care facility located in the Greater Accra Region. The current practice in effect has virtually cut-off infants in the other regions from accessing hearing screening and other pediatric audiological services. This has prompted a study into alternative methodologies to expand the reach of such services in Ghana. The present study was designed to assess the feasibility of using telehealth to deliver newborn hearing screening across Ghana.

**PURPOSE:** To assess the feasibility of using telehealth to extend newborn hearing screening services across the ten regions of Ghana.

**RESEARCH DESIGN:** A correlational study was designed to determine the extent of association between test results of telehealth and the conventional on-site methods (COMs) for conducting
newborn hearing screening. The design also allowed for testing duration between the two methods to be compared.

**STUDY SAMPLE:** Fifty infants from the Brong-Ahafo Regional Hospital (BARH) were enrolled. The infants aged between 2 and 90 days were selected through convenience sampling. There were 30 males and 20 females.

**PROCEDURE:** Newborn hearing screening using distortion product otoacoustic emissions were performed via telehealth. By adopting the synchronous telehealth model, an audiologist located at the Korle-Bu Teaching Hospital conducted real-time hearing screening tests over the internet on infants who were at the BARH. The former and latter hospitals are located in the Greater Accra and the Brong-Ahafo Regions, respectively. As a control, similar hearing screening tests were conducted on the same infants at BARH using the conventional face-to-face on-site hearing screening method.

**DATA COLLECTION AND ANALYSIS:** The test results and testing duration of the telehealth method and the conventional on-site approach were compared and subjected to statistical analysis. Here, the Spearman's correlation coefficient (rs) was used to determine the level of correlation between the test results, whereas the paired t-test statistic was used to test the level of significance between the testing duration of the two methods.

**RESULTS:** Analysis of the test results showed a significantly high positive correlation between the telehealth and the COMs (rs = 0.778, 0.878, 0.857, 0.823, p < 0.05). The mean testing duration (in seconds) of telehealth was 27.287 (standard deviation = 27.373) and that of the COM was 24.689 (standard deviation = 27.169).

**CONCLUSION:** The study showed the feasibility of establishing an interregional network of newborn hearing screening services across Ghana using telehealth. It is more efficient to deploy telehealth for pediatric hearing services than to have patients travel many hours to the Greater Accra Region for similar services. Poor road network, high transportation costs, and bad weather conditions are a few of the reasons for avoiding long distance travel in Ghana.

**Database:** Medline

**21. Sensorineural hearing loss in children with sickle cell disease.**

**Author(s):** Farrell, Annie N; Landry, April M; Yee, Marianne E; Leu, Roberta M; Goudy, Steven L

**Source:** International journal of pediatric otorhinolaryngology; Mar 2019; vol. 118; p. 110-114

**Publication Date:** Mar 2019

**Publication Type(s):** Journal Article

**PubMedID:** 30599285

**Abstract:**

**INTRODUCTION:** Sensorineural hearing loss (SNHL) has been reported to occur at increased frequency in the pediatric sickle cell disease (SCD) population, likely secondary to ototoxic medication regimens and repeat sickling events that lead to end organ damage. Risk and protective factors of SNHL in this population are not fully characterized. The objective of this study was to describe audiology results in children with SCD and the prevalence and sequelae of SNHL.

**METHODS:** A comprehensive clinical database of 2600 pediatric SCD patients treated at 1 institution from 2010-16 was retrospectively reviewed to identify all patients who were referred for audiologic testing. Audiologic test results, patient characteristics, and SCD treatments were reviewed.
RESULTS: 181 SCD children (97 male, 153 HbSS) underwent audiologic testing, with 276 total audiology encounters, ranging 1-9 per patient. Mean age at first audiogram was 8.9 ± 5.2 years. 29.8% had prior cerebrovascular infarct and an additional 25.4% had prior abnormal transcranial Doppler screens documented at time of first audiogram. Overall, 13.3% had documented hearing loss, with 6.6% SNHL. Mean pure tone average (PTA) among patients with SNHL ranged from mild to profound hearing loss (Right: 43.3 ± 28.9, Left: 40.8 ± 29.7), sloping to more severe hearing loss at higher frequencies.

CONCLUSIONS: Hearing loss was identified in a significant subset of children with SCD and the hearing loss ranged from normal to profound. Though the overall prevalence of SNHL in SCD patients was low, baseline audiology screening should be considered.

Database: Medline


Author(s): den Besten, Christine A; Monksfield, Peter; Bosman, Arjan; Skarzynski, Piotr H; Green, Kevin; Runge, Christina; Wigren, Stina; Blechert, Johan I; Flynn, Mark C; Mylanus, Emmanuel A M; Hol, Myrthe K S

Source: Clinical otolaryngology : official journal of ENT-UK ; official journal of Netherlands Society for Oto-Rhino-Laryngology & Cervico-Facial Surgery; Mar 2019; vol. 44 (no. 2); p. 144-157

Publication Date: Mar 2019
Publication Type(s): Journal Article
PubMedID: 30358920
Available at Clinical otolaryngology : official journal of ENT-UK ; official journal of Netherlands Society for Oto-Rhino-Laryngology & Cervico-Facial Surgery - from Wiley Online Library

Abstract:

OBJECTIVES: To compare the hearing performance of patients with conductive and mild mixed hearing loss and single-sided sensorineural deafness provided with a new transcutaneous bone conduction hearing implant (the Baha Attract System) with unaided hearing as well as aided with a sound processor on a softband. Furthermore, to evaluate safety and subjective benefit before and after implantation of the test device.

PARTICIPANTS: Fifty-four adult patients in five participating centres were enrolled in this prospective study. Baseline data were collected during a pre-operative visit, and after a softband trial, all patients were implanted unilaterally. Follow-up visits were scheduled at 10 days, 4, 6, 12 weeks and 6 months.

MAIN OUTCOME MEASURES: Free-field hearing thresholds pure-tone average (PTA4 in dB HL; mean threshold at 500, 1000, 2000, 4000 Hz; primary outcome measure). Individual free-field hearing thresholds, speech recognition in quiet and in noise, soft tissue status during follow-up and subjective benefit as measured with the Abbreviated Profile of Hearing Aid Benefit (APHAB), Speech, Spatial and Qualities of Hearing Scale (SSQ) and Health Utilities Index (HUI) questionnaires.

RESULTS: Implantation of the Baha Attract System resulted in favourable audiological outcomes compared to unaided conditions. On the primary outcome parameter, a statistically significant improvement was observed compared to unaided hearing for the patients with conductive/mixed hearing loss (mean PTA4 difference -20.8 dB HL, SD 9.8; P < 0.0001) and for the patients with single-
sided sensorineural deafness (SSD) (mean PTA4 difference -21.6 dB HL, SD 12.2; P 0.05). Soft tissue-related issues observed during follow-up included numbness, pain/discomfort at the implant site and to a lesser extent pressure-related skin complications. A declining trend was noted in the rate of these complications during follow-up. Approximately 20% of patients reported some degree of numbness and 38% (slight) pain/discomfort at final follow-up of 6 months. Good results on the subjective benefit questionnaires were observed, with statistically significant improvements on APHAB and SSQ questionnaires, and on the hearing attribute of HUI3.

**CONCLUSIONS:** The Baha Attract System provided a significant improvement in hearing performance and subjective benefit compared to the pre-operative unaided condition (with the non-test ear blocked). Hearing performance of the Baha Attract was similar to a test situation with the same sound processor on a softband. A proportion of the patients reported numbness and pain/discomfort at the implant site during follow-up, especially during the first post-operative weeks. Based on the results of the current multicentre study, the Baha Attract can be considered as a treatment option for patients with the aforementioned hearing losses. Especially in the SSD patients, a careful selection procedure is warranted. Therefore, a pre-operative trial should be part of the decision-making process before fitting a patient with the Baha Attract System.

**Database:** Medline


**Author(s):** Ramos Macías, Ángel; Borkoski-Barreiro, Silvia A; Falcón González, Juan C; de Miguel Martínez, Isabel; Ramos de Miguel, Ángel

**Source:** Clinical otolaryngology : official journal of ENT-UK ; official journal of Netherlands Society for Oto-Rhino-Laryngology & Cervico-Facial Surgery; Mar 2019; vol. 44 (no. 2); p. 138-143

**Publication Date:** Mar 2019

**Publication Type(s):** Journal Article

**PubMedID:** 30354002

Available at [Clinical otolaryngology : official journal of ENT-UK ; official journal of Netherlands Society for Oto-Rhino-Laryngology & Cervico-Facial Surgery](from Wiley Online Library)

**Abstract:**

**OBJECTIVE:** To determine the audiological and clinical results of cochlear implantation in children below the age of 12 years old with congenital and acquired single-sided deafness.

**DESIGN:** Observational, descriptive, transversal study.

**MAIN OUTCOME MEASURES:** Speech reception thresholds, Cortical responses, Auditory Lateralization Test and SSQ questionnaire.

**PARTICIPANTS:** Children < 12 implanted for congenital or acquired SSD.

**RESULTS:** All the children with congenital SSD showed positive cortical responses. Positive results were obtained in the Auditory Lateralization Test for the following modalities: 0º, 45º and 90º. With respect to the Speech Test, the children with acquired SSD showed the following results: 92% and 100% in recognition and 48% and 68% (Azimuth modalities), Signal CI side 52% and 68% and Signal normal hearing side 44% - 60% (p < 0.05). In both group the processor was used for 6-12 hours. With respect to the SSQ questionnaire results, the parents were more satisfied within the post-operative period than within the pre-operative period (P<0.001).
CONCLUSIONS: Cochlear implant provides children with congenital SSD with significant audiological and subjective benefits. Children with congenital SSD and implanted after a longer period may not have an important benefit (binaural) although other bilateral effects can be achieved. Children with post-lingual unilateral deafness and after a short period of hearing deprivation probably integrated the normal acoustic hearing with the cochlear implant electrical signal and showed binaural benefits.

Database: Medline

24. Hearing impairment after subarachnoid hemorrhage.

Author(s): Campbell, Nicci; Verschuur, Carl; Mitchell, Sophie; McCaffrey, Orlaith; Deane, Lewis; Taylor, Hannah; Smith, Rory; Foulkes, Lesley; Glazier, James; Darekar, Angela; Haacke, Mark E; Bulters, Diederik; Galea, Ian

Source: Annals of clinical and translational neurology; Mar 2019; vol. 6 (no. 3); p. 420-430

Publication Date: Mar 2019

Publication Type(s): Journal Article

PubMedID: 30911566

Available at Annals of clinical and translational neurology - from Europe PubMed Central - Open Access

Available at Annals of clinical and translational neurology - from Unpaywall

Abstract:

Background: Subarachnoid hemorrhage (SAH) survivors experience significant neurological disability, some of which is under-recognized by neurovascular clinical teams. We set out to objectively determine the occurrence of hearing impairment after SAH, characterize its peripheral and/or central origin, and investigate likely pathological correlates.

Methods: In a case-control study (n = 41), participants were asked about new onset hearing difficulty 3 months post-SAH, compared with pre-SAH. Formal audiological assessment included otoscopy, pure tone audiometry, a questionnaire identifying symptoms of peripheral hearing loss and/or auditory processing disorder, and a test of speech understanding in noise. A separate cohort (n = 21) underwent quantitative susceptibility mapping (QSM) of the auditory cortex 6 months after SAH, for correlation with hearing difficulty.

Results: Twenty three percent of SAH patients reported hearing difficulty that was new in onset post-SAH. SAH patients had poorer pure tone thresholds compared to controls. The proportion of patients with peripheral hearing loss as defined by the World Health Organization and British Audiological Society was however not increased, compared to controls. All SAH patients experienced symptoms of auditory processing disorder post-SAH, with speech-in-noise test scores significantly worse versus controls. Iron deposition in the auditory cortex was higher in patients reporting hearing difficulty versus those who did not.

Conclusion: This study firmly establishes hearing impairment as a frequent clinical feature after SAH. It primarily consists of an auditory processing disorder, mechanistically linked to iron deposition in the auditory cortex. Neurovascular teams should inquire about hearing, and refer SAH patients for audiological assessment and management.

Database: Medline
25. Tinnitus in the side with better hearing.

Author(s): Lee, Ho Yun; Kim, Su Jin; Chang, Dong Sik; Shin, Sun Ae
Source: American journal of otolaryngology; Feb 2019
Publication Date: Feb 2019
Publication Type(s): Journal Article
PubMedID: 30799211

Abstract:

OBJECTIVES: We aimed to confirm the characteristics of patients with tinnitus in the better-hearing side.

MATERIALS AND METHODS: Among the 778 patients who visited the tinnitus clinic complaining of unilateral tinnitus at a local university hospital between March 2014 and December 2017, we recruited 62 patients who showed tinnitus in the better-hearing side on pure-tone audiometry. The mean hearing threshold was calculated using the arithmetic mean of the pure tone thresholds at 1, 2, 3, and 4 kHz. In addition, patients' medical history, tinnitus questionnaires, and other audiological test results were thoroughly analyzed together for diagnosis.

RESULTS: Fluctuating hearing loss without vertigo or Ménière's disease were the most common etiologies (n = 16, 25.8%), followed by high-frequency hearing loss (n = 13, 21.0%), sudden idiopathic hearing loss (n = 6, 9.7%), and presbycusis (n = 6, 9.7%). Somatosensory tinnitus was also observed in seven patients. Neck pain was associated with tinnitus in five patients (8.1%), and two other patients (3.2%) experienced temporomandibular disorder in the same side as the tinnitus.

CONCLUSION: Tinnitus was associated with deterioration of hearing even when it occurred in the better-hearing side. Among the possible etiologies, fluctuating hearing loss in the tinnitus side was the most common audiologic finding. Assessment of hearing level at each frequency was more effective in detecting high-frequency hearing loss rather than the use of the mean hearing level. In addition, somatosensory tinnitus should not be ignored.

Database: Medline


Author(s): Lee, Jennifer W; Bance, Manohar L
Source: Practical neurology; Feb 2019; vol. 19 (no. 1); p. 28-35
Publication Date: Feb 2019
Publication Type(s): Journal Article
PubMedID: 30185631

Abstract: Hearing loss affects one in six people in the UK and is a significant disease burden. In addition to communication problems, there is also an association with depression and dementia. Clinical assessment with targeted history and examination can identify the characteristics and cause of hearing loss, and complementary audiological testing can confirm its type and severity. Retrocochlear screening is recommended for sudden, rapidly progressive or asymmetric sensorineural hearing loss. Medical or surgical therapies may be indicated in cases of conductive
hearing loss, while hearing assistive devices and hearing aids are the mainstay of rehabilitation for sensorineural hearing loss.

Database: Medline

27. Effects of oral zinc supplementation on patients with noise-induced hearing loss associated tinnitus: A clinical trial.

Author(s): Yeh, Chun-Wei; Tseng, Leng-Hsuan; Yang, Chao-Hui; Hwang, Chung-Feng

Source: Biomedical journal; Feb 2019; vol. 42 (no. 1); p. 46-52

Publication Date: Feb 2019

Publication Type(s): Journal Article

PubMedID: 30987704

Available at Biomedical journal - from ProQuest (Health Research Premium) - NHS Version

Abstract:

BACKGROUND: Zinc plays a vital antioxidant role in human metabolism. Recent studies have demonstrated a correlation between noise-induced hearing loss (NIHL) and oxidative injury; however, no investigation has focused specifically on the subgroup of NIHL associated tinnitus patients. We aimed to evaluate the effectiveness of zinc supplementation in treating NIHL associated tinnitus.

METHODS: Twenty patients with tinnitus and a typical NIHL audiogram (38 ears) were included in this study. Another 20 healthy subjects were used as the control group. A full medical history assessment was performed, and each subject underwent an otoscopic examination, basic audiolologic evaluation, distrotion product otoacoustic emissions (DPOAEs), tinnitus-match testing, Tinnitus Handicap Inventory (THI) and serum zinc level analyses. After 2 months of treatment with zinc, all tests were repeated.

RESULTS: There was a significant difference between pretreatment and post-treatment within the tinnitus group (73.6 vs. 84.6 μg/dl). The pre- and post-treatment difference in serum zinc was significantly higher in the young group (≦50 years) compared to the old group (19.4 ± 11.4 vs. 2.6 ± 9.2 μg/dl, respectively; p = 0.002). There were no statistically significant differences in hearing thresholds, speech reception thresholds, or tinnitus frequency and loudness results before and after treatment. In addition, 17 patients (85%) showed statistically significant improvement of THI-total scores post-treatment, from 38.3 to 30 (p = 0.024).

CONCLUSIONS: Zinc oral supplementation elevated serum zinc levels, especially in younger patients. THI scores improved significantly following zinc treatment in patients with NIHL associated tinnitus. However, no improvements in objective hearing parameters were observed.

Database: Medline


Author(s): Planey, Arrianna Marie

Source: Social science & medicine (1982); Jan 2019; vol. 222 ; p. 216-224

Publication Date: Jan 2019
This study employs statistical modeling and mapping techniques to analyze the availability and accessibility of audiologists (practitioners who diagnose and treat hearing loss) in the United States at the county scale. The goal is to assess the relationships between socio-demographic and structural factors (such as health policy and clinical programs which train audiologists) and audiologist availability. These associations are analyzed at the county level, via a mixed effects hurdle model. At the county level, the proportion of older adults reporting difficulty hearing is negatively associated with audiologist supply. The findings show that audiologists tend to locate in metropolitan counties with higher median household incomes, younger populations, and lower proportions of older adults reporting hearing difficulty, suggesting an inverse care-type relationship between audiologist availability and need for hearing health services. Notably, neither state legislation requiring insurance plan coverage of hearing services for adults or Medicaid coverage of audiology services were significant predictors of audiologist supply at the county level.

Database: Medline

29. Assessing and managing concurrent hearing, vision and cognitive impairments in older people: an international perspective from healthcare professionals.

Author(s): Leroi, Iracema; Himmelsbach, Ines; Wolski, Lucas; Littlejohn, Jenna; Jury, Francine; Parker, Angela; Charalambous, Anna Pavlina; Dawes, Piers; Constantinidou, Fofi; Thodi, Chryssoula; (SENSE-Cog Expert Reference Group)

Source: Age and ageing; Jan 2019

Publication Date: Jan 2019

Publication Type(s): Journal Article

Abstract:

Background: there is a significant gap in the understanding, assessment and management of people with dementia and concurrent hearing and vision impairments.

Objective: from the perspective of professionals in dementia, hearing and vision care, we aimed to: (1) explore the perceptions of gaps in assessment and service provision in ageing-related hearing, vision and cognitive impairment; (2) consider potential solutions regarding this overlap and (3) ascertain the attitudes, awareness and practice, with a view to implementing change.

Methods: our two-part investigation with hearing, vision, and dementia care professionals involved: (1) an in-depth, interdisciplinary, international Expert Reference Group (ERG; n = 17) and (2) a wide-scale knowledge, attitudes and practice survey (n = 653). The ERG involved consensus discussions around prototypic clinical vignettes drawn from a memory centre, an audiology clinic, and an optometry clinic, analysed using an applied content approach.

Results: the ERG revealed several gaps in assessment and service provision, including a lack of validated assessment tools for concurrent impairments, poor interdisciplinary communication and care pathways, and a lack of evidence-based interventions. Consensus centred on the need for flexible, individualised, patient-centred solutions, using an interdisciplinary approach. The survey data validated these findings, highlighting the need for clear guidelines for assessing and managing concurrent impairments.
Conclusions: this is the first international study exploring professionals’ views of the assessment and care of individuals with age-related hearing, vision and hearing impairment. The findings will inform the adaptation of assessments, the development of supportive interventions, and the new provision of services.

Database: Medline


Author(s): Ramkumar, Vidya; Nagarajan, Roopa; Shankarnarayan, Vanaja C; Kumaravelu, Selvakumar; Hall, James W

Source: BMC health services research; Jan 2019; vol. 19 (no. 1); p. 1

Publication Date: Jan 2019

Publication Type(s): Journal Article

PubMedID: 30606168

Available at BMC health services research - from BioMed Central

Available at BMC health services research - from Europe PubMed Central - Open Access

Available at BMC health services research - from EBSCO (MEDLINE Complete)

Available at BMC health services research - from ProQuest (Health Research Premium) - NHS Version

Available at BMC health services research - from Unpaywall

Abstract:

BACKGROUND: In an attempt to reach remote rural areas, this study explores a community-based, pediatric hearing screening program in villages, integrating two models of diagnostic ABR testing; one using a tele-medicine approach and the other a traditional in-person testing at a tertiary care hospital.

METHODS: Village health workers (VHWs) underwent a five day training program on conducting Distortion Product Oto Acoustic Emissions (DPOAE) screening and assisting in tele-ABR. VHWs conducted DPOAE screening in 91 villages and hamlets in two administrative units (blocks) of a district in South India. A two-step DPOAE screening was carried out by VHWs in the homes of infants and children under five years of age in the selected villages. Those with ‘refer’ results in 2nd screening were recommended for a follow-up diagnostic ABR testing in person (Group A) at the tertiary care hospital or via tele-medicine (Group B). The overall outcome of the community-based hearing screening program was analyzed with respect to coverage, refer rate, follow-up rate for 2nd screenings and diagnostic testing. A comparison of the outcomes of tele-versus in-person diagnostic ABR follow-up was carried out.

RESULTS: Six VHWs who fulfilled the post training evaluation criteria were recruited for the screening program. VHWs screened 1335 children in Group A and 1480 children in Group B. The refer rate for 2nd screening was very low (0.8%); the follow-up rate for 2nd screening was between 80 and 97% across the different age groups. Integration of tele-ABR resulted in 11% improvement in follow-up compared to in-person ABR at a tertiary care hospital.

CONCLUSIONS: Non-availability of audiologists and limited infrastructure in rural areas has prevented the establishment of large scale hearing screening programs. In existing programs, considerable challenges with respect to follow-up for diagnostic testing was reported, due to
patients being submitted to traveling long distance to access services and potential wage losses during that time. In this program model, integration of a tele-ABR diagnostic follow-up improved follow-up in comparison to in-person follow-up. VHWs were successfully trained to conduct accurate screenings in rural communities. The very low refer rate, and improved follow-up rate reflect the success of this community-based hearing screening program.

**Database:** Medline

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**31. Speech-in-noise representation in the aging midbrain and cortex: Effects of hearing loss.**

**Author(s):** Presacco, Alessandro; Simon, Jonathan Z; Anderson, Samira

**Source:** PloS one; 2019; vol. 14 (no. 3); p. e0213899

**Publication Date:** 2019

**Publication Type(s):** Journal Article

**PubMedID:** 30865718

*Available at:* PloS one - from Europe PubMed Central - Open Access

*Available at:* PloS one - from Public Library of Science (PLoS)

*Available at:* PloS one - from EBSCO (MEDLINE Complete)

*Available at:* PloS one - from Unpaywall

**Abstract:** Age-related deficits in speech-in-noise understanding pose a significant problem for older adults. Despite the vast number of studies conducted to investigate the neural mechanisms responsible for these communication difficulties, the role of central auditory deficits, beyond peripheral hearing loss, remains unclear. The current study builds upon our previous work that investigated the effect of aging on normal-hearing individuals and aims to estimate the effect of peripheral hearing loss on the representation of speech in noise in two critical regions of the aging auditory pathway: the midbrain and cortex. Data from 14 hearing-impaired older adults were added to a previously published dataset of 17 normal-hearing younger adults and 15 normal-hearing older adults. The midbrain response, measured by the frequency-following response (FFR), and the cortical response, measured with the magnetoencephalography (MEG) response, were recorded from subjects listening to speech in quiet and noise conditions at four signal-to-noise ratios (SNRs): +3, 0, -3, and -6 dB sound pressure level (SPL). Both groups of older listeners showed weaker midbrain response amplitudes and overrepresentation of cortical responses compared to younger listeners. No significant differences were found between the two older groups when the midbrain and cortical measurements were analyzed independently. However, significant differences between the older groups were found when investigating the midbrain-cortex relationships; that is, only hearing-impaired older adults showed significant correlations between midbrain and cortical measurements, suggesting that hearing loss may alter reciprocal connections between lower and higher levels of the auditory pathway. The overall paucity of differences in midbrain or cortical responses between the two older groups suggests that age-related temporal processing deficits may contribute to older adults' communication difficulties beyond what might be predicted from peripheral hearing loss alone; however, hearing loss does seem to alter the connectivity between midbrain and cortex. These results may have important ramifications for the field of audiology, as it indicates that algorithms in clinical devices, such as hearing aids, should consider age-related temporal processing deficits to maximize user benefit.

**Database:** Medline

**Author(s):** Bois, Emilie; Francois, Martine; Benkerrou, Malika; Van Den Abbeele, Thierry; Teissier, Natacha

**Source:** Pediatric blood & cancer; Jan 2019; vol. 66 (no. 1); p. e27468

**Publication Date:** Jan 2019

**Publication Type(s):** Journal Article

**PubMedID:** 30251366

Available at [Pediatric Blood & Cancer](https://onlinelibrary.wiley.com/journal/10.1002) - from Wiley Online Library

**Abstract:**

**BACKGROUND:** Sickle cell disease (SCD) is the most common genetic disease in France. In developing countries, it is associated with a high incidence of hearing loss. The aim of this study was to determine the prevalence of hearing loss in French children with SCD in order to determine if they need a close audiological follow-up.

**METHODS:** We performed a single-center prospective cross-sectional study of children with SCD. The children, without specific hearing symptom, underwent an ear, nose and throat examination with a hearing assessment between 2015 and 2016.

**RESULTS:** Eighty-nine children were included, aged from 5 to 19 years, with 73% of SS or Sβ0 genotype and 27% of SC or Sβ+ genotype. Ten children (11.2%) had hearing thresholds higher than 20 dB in at least one ear: one child with subnormal hearing, six otitis media with effusion (OME), and three sensorineural hearing loss. Late age at diagnosis of SCD, a high platelet count and a low hematocrit level were significantly associated with OME; moreover, children with OME had more severe clinical and biological characteristics than children with normal hearing. Furthermore, 12.4% of the children complained of tinnitus. The rate of sudden hearing loss was 2.2%. Finally, 7.1% of patients with normal hearing showed a speech discrimination disorder.

**CONCLUSIONS:** Several causes were identified for hearing loss in children with SCD. They therefore need a close audiological follow-up in order to avoid complications due to curable phenomena and to enable appropriate management for progressive complications.

**Database:** Medline

33. Hearing Status in Survivors of Childhood Acute Myeloid Leukemia Treated With Chemotherapy Only: A NOPHO-AML Study.

**Author(s):** Skou, Anne-Sofie; Olsen, Steen Ø; Nielsen, Lars H; Glosli, Heidi; Jahnikainen, Kirsi; Jarfelt, Marianne; Jónmundsson, Guðmundur K; Malmros, Johan; Nysom, Karsten; Hasle, Henrik; Nordic Society of Pediatric Hematology and Oncology (NOPHO)

**Source:** Journal of pediatric hematology/oncology; Jan 2019; vol. 41 (no. 1); p. e12

**Publication Date:** Jan 2019

**Publication Type(s):** Journal Article

**PubMedID:** 30550508

**Abstract:**
BACKGROUND: As more children survive acute myeloid leukemia (AML) it is increasingly important to assess possible late effects of the intensive treatment. Hearing loss has only sporadically been reported in survivors of childhood AML. We assessed hearing status in survivors of childhood AML treated with chemotherapy alone according to 3 consecutive NOPHO-AML trials.

PROCEDURE: A population-based cohort of children treated according to the NOPHO-AML-84, NOPHO-AML-88, and NOPHO-AML-93 trials included 137 eligible survivors among whom 101 (74%) completed a questionnaire and 99 (72%) had otologic and audiologic examination performed including otoscopy (72%), pure tone audiometry (70%), and tympanometry (60%). Eighty-four of 93 (90%) eligible sibling controls completed a similar questionnaire.

RESULTS: At a median of 11 years (range, 4 to 25) after diagnosis, hearing disorders were rare in survivors of childhood AML and in sibling controls, with no significant differences. None had severe or profound hearing loss diagnosed at audiometry. Audiometry detected a subclinical hearing loss ranging from slight to moderate in 19% of the survivors, 5% had low-frequency hearing loss, and 17% had high-frequency hearing loss.

CONCLUSIONS: The frequency of hearing disorders was low, and hearing thresholds in survivors of childhood AML were similar to background populations of comparable age.

Database: Medline

34. Hidden hearing loss in children and adolescents with sickle cell anemia.

Author(s): Rissatto-Lago, Mara Renata; da Cruz Fernandes, Luciene; Lyra, Isa Menezes; Terse-Ramos, Regina; Teixeira, Rozana; Salles, Cristina; Teixeira Ladeia, Ana Marice

Source: International journal of pediatric otorhinolaryngology; Jan 2019; vol. 116 ; p. 186-191

Publication Date: Jan 2019

Publication Type(s): Journal Article

PubMedID: 30554696

Abstract:

OBJECTIVE: To evaluate the auditory system for hidden hearing loss (HHL) and its association with clinical variables and endothelial dysfunction (ED) in children and adolescents with sickle cell anemia (SCA).

METHODS: Participants included 37 patients with stable SCA and 44 healthy controls (HC group) (aged 6-18 years) with hearing thresholds ≤ 20 dB (dB) were evaluated for pure tone audiometry, tympanometry, acoustic reflex, otoacoustic emission, and auditory evoked potentials. Laboratory analysis of the lipid profile, and C-reactive protein levels and endothelial function using ultrasonographic imaging of the brachial artery to assess flow-mediated dilation were performed.

RESULTS: The SCA group presented with a higher rate of increased contralateral acoustic reflex thresholds, compared to those in the HC group at all frequencies and in both ears (p < 0.05). There were significant differences in the brainstem auditory evoked potentials between the SCA and HC groups. In the SCA group, the waves III and V latencies were increased (p = 0.006 and 0.004 respectively), and the I-III and I-V interpeak intervals were longer (p = 0.015 and 0.018 respectively) than those in the HC group. There was no association between the audiological measures and clinical and metabolic variables and sickle cell anemia complications including endothelial function and therapy.
CONCLUSION: In conclusion, our findings suggest that damage in the auditory system in SCA patients can be present involving retrocochlear structures, causing functional deficits without deterioration of auditory sensitivity.

Database: Medline

35. Genotyping and audiological characteristics of infants with a single-allele SLC26A4 mutation.

Author(s): Zhao, Xuelei; Huang, Lihui; Wang, Xueyao; Wang, Xianlei; Zhao, Liping; Cheng, Xiaohua; Ruan, Yu

Source: International journal of pediatric otolorhinolaryngology; Jan 2019; vol. 116; p. 153-158

Publication Date: Jan 2019

Publication Type(s): Journal Article

PubMedID: 30554688

Abstract:

OBJECTIVES: To identify second-allele variant in infants with a known single-allele mutation of the SLC26A4 gene and to determine the frequency of their occurrence; and to investigate the clinical audiological characteristics of infants with bi-allelic mutations in SLC26A4.

METHODS: The study subjects were 371 patients with a single-allele SLC26A4 mutation detected by neonatal deafness gene screening (4 genes and 9 pathogenic variants) who were treated at the otology outpatient department of Beijing Tongren Hospital. The exonic and flanking splice site regions of the SLC26A4 gene were sequenced for all patients. All patients with bi-allelic SLC26A4 mutations underwent audiological evaluation, and some also underwent temporal bone computed tomography and/or inner ear magnetic resonance imaging.

RESULTS: Of the 371 patients, 314 (84.64%) had an c.919-2A > G heterozygous mutation and 57 (15.36%) had a c.2168A > G (p.H723R) heterozygous mutation. 13 patients (3.50%) had a second-allele variant, including 11 (2.96%) with pathogenic mutations and 1 (0.27%) with a likely benign variant. Of the 13 patients with bi-allelic mutations, 11 had hearing loss and 2 had normal hearing, the latter of whom had c.919-2A > G/c.1766A > G and c.919-2A > G/c.757A > G compound heterozygous mutations, respectively. Four of the 13 patients with bi-allelic mutations had passed the universal newborn hearing screening, including 2 cases (15.38%) with hearing loss. The most prevalent degree of hearing loss was profound (40.91%), followed by severe (36.36%). The most prevalent audiometric configuration was sloping hearing loss (50.00%), followed by flat-type hearing loss (40.91%).

CONCLUSIONS: This is the first report in China of the frequency of occurrence of second-allele variant in infants with a known single-allele mutation of the SLC26A4 gene; the frequency was 3.50% for any type of variant and 2.96% for pathogenic mutations. A novel variant, c.1766A > G (p.Q589R), which is likely benign, was identified. The pathogenicity of c.757A > G (p.I253V) mutation deserves more in-depth research. For infants with bi-allelic SLC26A4 mutations, the degree of hearing loss was mainly severe-to-profound and the audiometric configuration was mainly sloping.

Database: Medline

36. Otosyphilis: Resurgence of an Old Disease.

Author(s): Theeuwen, Hailey; Whipple, Mark; Litvack, Jamie R
OBJECTIVES: To describe the clinical characteristics of patients presenting with a new diagnosis of otosyphilis over the past 10 years in a large, urban, safety-net hospital affiliated with a large county sexually transmitted disease clinic.

METHODS: Retrospective case series. A chart review was performed of all patients who presented to an adult otolaryngology clinic with a new diagnosis of syphilis and hearing loss from January 2008 to December 2017.

RESULTS: Twelve patients met the criteria for "suspected" or "likely" otosyphilis based on Centers for Disease Control and Prevention definitions. The average age was 48 years (range 19-59). All were male. Nine (75%) were men who have sex with men. Eight (67%) were positive for human immunodeficiency virus. One (8%) presented with primary, nine (75%) with secondary, and two (17%) with early latent syphilis. Seven (58%) presented with bilateral audiogram-confirmed hearing loss, two (17%) with unilateral hearing loss, and three (25%) with suspected hearing loss based on fluctuating symptoms. Nine (75%) presented with tinnitus and two (17%) with vertigo. The median duration of otologic symptoms prior to presentation was 2 weeks (range: 0-16 weeks). All presented within the last 2 years surveyed.

CONCLUSION: We have seen an increase in the number of otosyphilis cases in our clinic. We suspect otosyphilis may be underdiagnosed and emphasize the importance of screening for syphilis in patients with new audiologic symptoms of vertigo, tinnitus, or hearing loss.LEVEL OF EVIDENCE4.

Database: Medline


Author(s): Boyce, Jessica O; Kilpatrick, Nicky; Teixeira, Rodrigo P; Morgan, Angela T

Source: Archives of disease in childhood. Education and practice edition; Dec 2018

Publication Date: Dec 2018

Publication Type(s): Journal Article

PubMedID: 30567832

Available at Archives of disease in childhood. Education and practice edition - from BMJ Journals

Abstract:

CASE HISTORY: A 5-year-old girl presented with a suspected palatal abnormality, first observed by her general practitioner during a routine examination at 4 years. She had a history of conductive hearing loss on a background of recurrent otitis media, however her hearing had improved since grommet insertion at 4 years and 8 months. Her middle ear dysfunction was thought to be due to abnormal palatal muscles that also control Eustachian tube drainage. No difficulties with breast feeding or nasal regurgitation were reported. She was referred to a cleft specialist team to be assessed by a cleft surgeon, speech language therapist and ear, nose and throat specialist. On oral
examination, she presented with a bifid uvula at rest (figure 1). An overt submucous cleft palate (SMCP) was confirmed through palpation and inspection of her palate during phonation of ‘ahh’ and ‘ah-ahh’, which allowed for visualisation of soft palate elevation. Speech assessment revealed typical resonance (ie, no hypernasality) and no nasal air emission or turbulence, the presence of which may typically suggest an SMCP. The patient’s speech was ‘often hard to understand’ however, but this was due to delayed phonology (cognitive-linguistic speech sound processing). She also had impaired receptive (understanding) and expressive (talking) language skills on formal standardised language assessment (Clinical Evaluation of Language Fundamentals, Fourth Edition).

Surgical intervention was not indicated given the cleft was not functionally impacting her speech (ie, no velopharyngeal insufficiency (VPI)) or eating (ie, no nasal regurgitation during feeding). Surgery aims to improve velopharyngeal functioning by repositioning the levator veli palatini muscle. This child had adequate velopharyngeal functioning despite her striking anatomical presentation, and her speech difficulties were developmental (phonological impairment) rather than structural. Speech therapy is indicated for her delayed phonology and language deficits. VPI symptoms will be monitored for late onset as she continues to grow. This is particularly important as children with palatal anomalies have increased risk of developing VPI when their adenoids atrophy with age. She will also remain under the care of ear, nose and throat specialists and audiologists to monitor her hearing and middle ear dysfunction.

QUESTIONS: What anatomical features indicate an overt SMCP? Any or all of the above What features of communication would indicate VPI and possible surgical intervention? High-pitched or rough sounding voice

Database: Medline

38. Auditory Evoked Potential Inconsistency in Sudden Unilateral Hearing Loss with Multiple Sclerosis.

Author(s): Lee, Sungsu; Jeon, Eun-Sun; Cho, Hyong-Ho

Source: The journal of international advanced otology; Dec 2018

Publication Date: Dec 2018

Publication Type(s): Journal Article

PubMedID: 30541734

Available at The journal of international advanced otology - from ProQuest (Health Research Premium) - NHS Version

Available at The journal of international advanced otology - from Unpaywall

Abstract: Sudden sensorineural hearing loss is a well-recognized clinical symptom in multiple sclerosis (MS). Acute inflammatory demyelination in the cochlear nerve or more central auditory tracts may cause sudden retrocochlear hearing loss. A 28-year-old male patient who was confirmed as having MS presented with suffering from dizziness as well as ongoing right-side hearing loss. We performed audiological tests, such as pure tone audiometry (PTA), otoacoustic emission, auditory brainstem response (ABR), and auditory steady-state response (ASSR). His clinical and audiological abnormalities disappeared with steroid therapy. However, each test showed different time courses.
of improvement. Although the results of the PTA and ASSR tests improved in exactly 1 month after the first attack, the results of the ABR reached 3 months to return to normal. To the best of our knowledge, this is the first case report of the time difference of hearing improvement shown in PTA, ASSR, and ABR tests.

Database: Medline

39. The evaluation of the sense of hearing in patients with carotid artery stenosis within the extracranial segments.

Author(s): Dorobisz, Karolina; Dorobisz, Tadeusz; Janczak, Dariusz; Krecicki, Tomasz

Source: Acta neurologica Belgica; Dec 2018

Publication Date: Dec 2018

Publication Type(s): Journal Article

PubMedID: 30542965

Abstract: Stenosis of arteries that supplies blood to the brain is one of the main causes of ischemic stroke which is the third most common cause of deaths in Europe. Atherosclerosis of carotid and vertebral arteries is responsible for 20% of the ischemic stroke cases. Stenosis may be either asymptomatic or manifested with typical neurological symptoms including motor and sensory disturbances as well as disturbances in vision and speech. However, discrete non-specific symptoms of ischemia, including headaches and vertigo, tinnitus and hearing loss, are also quite common. These symptoms may be indicative of a clinically significant stenosis of carotid and vertebral arteries, particularly within the internal carotid artery region, as well as of a risk of ischemic stroke. To date, research reports were unable to provide exact explanation of correlations between impaired hearing and the stenosis of carotid and vertebral arteries. Despite this, numerous articles list these symptoms as one of the first non-specific symptoms of this disorder. The ischemic mechanism within the inner ear region may lead to early symptoms of atherosclerosis of large vessels. However, no evidence of relationship and no explanation could be provided with this regard. The objective of the study was to assess the effect of carotid and vertebral artery stenosis on the function of the hearing and equilibrium organ on the basis of diagnostic audiological examinations including pure-tone threshold audiometry, impedance audiometry, otoacoustic emission tests and brainstem auditory evoked potential tests. The study was conducted in 63 patients (32 males, 31 females) aged 45-75 years, presenting with carotid and vertebral artery stenosis and treated at the Vascular Surgery Clinic of the University Clinical Hospital in Wroclaw. Patients were stratified into two subgroups according to their age (45-60 years, 61-75 years). Patients were also divided into subgroups according to the stenotic arteries and to the symptomatic/asymptomatic status of the disorder. All patients were homogeneous in terms of the degree of artery stenosis. The control group consisted of 32 healthy persons (14 males, 18 females) aged 48-75 years. Patients qualified to the control group reported no history of middle or inner ear disorders, disturbed hearing, vertigo and balance disorders, as well as cardiovascular diseases; they also presented with unremarkable ultrasound scans of the arteries. All patients were subjected to precise audiological examinations. Prior to being qualified for the study, patients were subjected to the assessment of arteries by means of Doppler ultrasonography. The hearing organ function was assessed by means of pure-tone threshold audiometry, impedance audiometry, otoacoustic emission tests and brainstem auditory evoked potential tests. Reduction of the flow through the carotid arteries causes problems in the organ of hearing; abnormalities are reported especially in tone threshold audiometry, examinations of the stapedius muscle reflexes and brainstem auditory evoked potentials, which prove the presence of
receptive cochlear-extracochlear hearing damage. Disturbances of the organ of hearing have similar severity in stenosis of the internal carotid artery and vertebral artery. Abnormalities found in audiologic examinations in patients with carotid artery stenosis are not always explicitly clinically expressed in patients with hearing loss; we should consider diagnostics for carotid artery stenosis.

**Database:** Medline

40. Congenital cytomegalovirus infection inducing non-congenital sensorineural hearing loss during childhood; a systematic review.

**Author(s):** Riga, Maria; Korres, George; Chouridis, Pantelis; Naxakis, Stephanos; Danielides, Vasilios

**Source:** International journal of pediatric otolaryngology; Dec 2018; vol. 115; p. 156-164

**Publication Date:** Dec 2018

**Publication Type(s):** Journal Article Review

**PubMedID:** 30368378

**Abstract:**

**BACKGROUND:** Congenital cytomegalovirus (CMV) infection is one of the most important risk factors for delayed onset and progressive hearing loss in children. However, the relevant literature is limited, heterogeneous and currently insufficient to provide guidance toward the effective monitoring of hearing acuity in these children.

**OBJECTIVES:** The aim of this study was to provide a systematic review focused on types of hearing loss that may escape diagnosis through universal neonatal hearing screening and/or present significant changes during childhood, such as progressive, fluctuating and late-onset hearing loss.

**DATA SOURCES:** A review of the present literature was conducted via the PubMed database of the US National Library of Medicine (www.pubmed.org) and Scopus database (www.scopus.com) with the search terms "late-onset hearing loss cytomegalovirus", "progressive hearing loss cytomegalovirus" and "fluctuating hearing loss cytomegalovirus".

**STUDY ELIGIBILITY CRITERIA:** Prospective or retrospective clinical studies were included if they presented a detailed audiological assessment, for a follow-up period of >2 years. METHODS: The prevalence and time of diagnosis of progressive, fluctuating and late-onset hearing loss were considered as primary outcomes. Results were recorded separately for symptomatic and asymptomatic children, when possible.

**RESULTS:** This analysis refers to a population of 181 children with CMV-induced hearing loss, who were diagnosed among 1089 with congenital CMV infection. The prevalence of CMV-induced hearing loss was significantly higher among symptomatic children (p < 0.0001), who were also significantly more likely to develop bilateral hearing loss (p = 0.001). There was not sufficient information on the prevalence, laterality, degree and time of diagnosis of progressive, fluctuating and late-onset hearing loss that could constitute the basis toward the report of specific follow-up guidelines.

**CONCLUSIONS:** Further studies are needed in order to understand and quantify the potential effects of congenital CMV infection in the inner ear and hearing acuity. The results presented in the relative studies should be very carefully evaluated and compared to each other, since they correspond to substantially different cohorts, study designs, and result elaboration. Infants with congenital CMV infection should be closely monitored, regarding their hearing acuity at least during their preschool years, although substantial changes in hearing thresholds have been reported as late as the 16th
year of age. Parental counseling is of outmost importance in order to minimize the numbers of children lost to follow-up.

Database: Medline

41. Predicting sequential bilateral cochlear implantation performance in postlingually deafened adults; A retrospective cohort study.

Author(s): Smulders, Yvette E; Hendriks, Thomas; Stegeman, Inge; Eikelboom, Robert H; Sucher, Cathy; Upson, Gemma; Chester Browne, Ronel; Jayakody, Dona; Santa Maria, Peter L; Atlas, Marcus D; Friedland, Peter L

Source: Clinical otolaryngology : official journal of ENT-UK ; official journal of Netherlands Society for Oto-Rhino-Laryngology & Cervico-Facial Surgery; Dec 2018; vol. 43 (no. 6); p. 1500-1507

Publication Date: Dec 2018

Publication Type(s): Journal Article

PubMedID: 30022607

Abstract:

OBJECTIVE: To identify which preoperative patient characteristics influence sequential bilateral cochlear implantation performance and to create a statistical model that predicts benefit.

DESIGN: Multicentre retrospective cohort study.

SETTING: All patients were operated in four academic teaching hospitals in Perth, Australia, and followed up by audiologists of the Ear Science Institute Australia.

PARTICIPANTS: A total of 92 postlingually deafened adult patients who had undergone sequential cochlear implantations between 19 June 1990 and 14 March 2016 were included. Patients were excluded if the 12-month follow-up consonant-nucleus-consonant (CNC) phoneme score was missing.

MAIN OUTCOME MEASURE: The effect of 18 preoperative factors on the CNC phoneme score in quiet (at 65 dB SPL) with the second cochlear implant (CI2) one year after implantation.

RESULTS: Two factors were positively correlated to speech understanding with CI2: Wearing a hearing aid (HA) before receiving CI2 (r = 0.46, P = 0.00) and the maximum CNC phoneme score with the first CI (CI1) (r = 0.21, P = 0.05). Two factors were negatively correlated: the length of hearing loss before CI2 in the second implanted ear (r = -0.25, P = 0.02) and preoperative pure tone average (PTA) (0.5, 1, 2 kHz) before CI2 in the second implanted ear (r = -0.27, P = 0.01). The following model could be created: predicted CNC phoneme score with CI2 (%) = 16 + (44 * HA use before CI2 (yes)) - (0.22 * length of hearing loss before CI2 (years)) + (0.23 * CNC phoneme score with CI1 (%)). Because the effect of HA use before implantation played such a major role, we also created a model after exclusion of the HA factor: Predicted CNC phoneme score with CI2 (%) = 82 - (0.17 * length of hearing loss before CI2 (years)) - (0.27 * PTA in second implanted ear before CI2 (0.5, 1, 2 kHz)) + (0.20 * CNC phoneme score with CI1 (%)).

CONCLUSION: Advanced age or a long interval between implantations does not necessarily lead to poor CI2 results. Patients who are successful HA users before CI2, who have a low PTA before CI2,
high CNC phoneme score with CI1 and a limited length of hearing loss before CI2, are likely to be successful CI2 recipients.

Database: Medline

42. Sound therapy (using amplification devices and/or sound generators) for tinnitus.

Author(s): Sereda, Magdalena; Xia, Jun; El Refaie, Amr; Hall, Deborah A; Hoare, Derek J

Source: The Cochrane database of systematic reviews; Dec 2018; vol. 12 ; p. CD013094

Publication Date: Dec 2018

Publication Type(s): Research Support, Non-u.s. Gov't Meta-analysis Journal Article Systematic Review

PubMedID: 30589445

Available at The Cochrane database of systematic reviews - from Cochrane Collaboration (Wiley)

Abstract:

BACKGROUND: Tinnitus affects 10% to 15% of the adult population, with about 20% of these experiencing symptoms that negatively affect quality of life. In England alone there are an estimated ¾ million general practice consultations every year where the primary complaint is tinnitus, equating to a major burden on healthcare services. Clinical management strategies include education and advice, relaxation therapy, tinnitus retraining therapy (TRT), cognitive behavioural therapy (CBT), sound enrichment using ear-level sound generators or hearing aids, and drug therapies to manage co-morbid symptoms such as insomnia, anxiety or depression. Hearing aids, sound generators and combination devices (amplification and sound generation within one device) are a component of many tinnitus management programmes and together with information and advice are a first line of management in audiology departments for someone who has tinnitus.

OBJECTIVES: To assess the effects of sound therapy (using amplification devices and/or sound generators) for tinnitus in adults.

SEARCH METHODS: The Cochrane ENT Information Specialist searched the Cochrane ENT Register; Central Register of Controlled Trials (CENTRAL, via the Cochrane Register of Studies); Ovid MEDLINE; Ovid Embase; CINAHL; Web of Science; ClinicalTrials.gov; ICTRP and additional sources for published and unpublished trials. The date of the search was 23 July 2018.

SELECTION CRITERIA: Randomised controlled trials (RCTs) recruiting adults with acute or chronic subjective idiopathic tinnitus. We included studies where the intervention involved hearing aids, sound generators or combination hearing aids and compared them to waiting list control, placebo or education/information only with no device. We also included studies comparing hearing aids to sound generators, combination hearing aids to hearing aids, and combination hearing aids to sound generators.

DATA COLLECTION AND ANALYSIS: We used the standard methodological procedures expected by Cochrane. Our primary outcomes were tinnitus symptom severity as measured as a global score on multi-item tinnitus questionnaire and significant adverse effects as indicated by an increase in self-reported tinnitus loudness. Our secondary outcomes were depressive symptoms, symptoms of generalised anxiety, health-related quality of life and adverse effects associated with wearing the device such as pain, discomfort, tenderness or skin irritation, or ear infections. We used GRADE to assess the quality of evidence for each outcome; this is indicated in italics.
MAIN RESULTS: This review included eight studies (with a total of 590 participants). Seven studies investigated the effects of hearing aids, four combination hearing aids and three sound generators. Seven studies were parallel-group RCTs and one had a cross-over design. In general, risk of bias was unclear due to lack of detail about sequence generation and allocation concealment. There was also little or no use of blinding. No data for our outcomes were available for any of our three main comparisons (comparing hearing aids, sound generators and combination devices with a waiting list control group, placebo or education/information only). Data for our additional comparisons (comparing these devices with each other) were also few, with limited potential for data pooling. Hearing aid only versus sound generator device only: One study compared patients fitted with sound generators versus those fitted with hearing aids and found no difference between them in their effects on our primary outcome, tinnitus symptom severity measured with the Tinnitus Handicap Inventory (THI) at 3, 6 or 12 months (low-quality evidence). The use of both types of device was associated with a clinically significant reduction in tinnitus symptom severity. Combination hearing aid versus hearing aid only: Three studies compared combination hearing aids with hearing aids and measured tinnitus symptom severity using the THI or Tinnitus Functional Index. When we pooled the data we found no difference between them (standardised mean difference -0.15, 95% confidence interval -0.52 to 0.22; three studies; 114 participants) (low-quality evidence). The use of both types of device was again associated with a clinically significant reduction in tinnitus symptom severity. Adverse effects were not assessed in any of the included studies. None of the studies measured the secondary outcomes of depressive symptoms or depression, anxiety symptoms or generalised anxiety, or health-related quality of life as measured by a validated instrument, nor the newly developed core outcomes tinnitus intrusiveness, ability to ignore, concentration, quality of sleep and sense of control.

AUTHORS’ CONCLUSIONS: There is no evidence to support the superiority of sound therapy for tinnitus over waiting list control, placebo or education/information with no device. There is insufficient evidence to support the superiority or inferiority of any of the sound therapy options (hearing aid, sound generator or combination hearing aid) over each other. The quality of evidence for the reported outcomes, assessed using GRADE, was low. Using a combination device, hearing aid or sound generator might result in little or no difference in tinnitus symptom severity. Future research into the effectiveness of sound therapy in patients with tinnitus should use rigorous methodology. Randomisation and blinding should be of the highest quality, given the subjective nature of tinnitus and the strong likelihood of a placebo response. The CONSORT statement should be used in the design and reporting of future studies. We also recommend the use of validated, patient-centred outcome measures for research in the field of tinnitus.

Database: Medline
NICE Updates:

Cochlear implants for children and adults with severe to profound deafness
Technology appraisal guidance [TA566]
Published: 07 March 2019
https://www.nice.org.uk/guidance/ta566

subsequently the NICE pathway has been updated

Hearing loss overview
NICE Pathway
Last updated: March 2019
https://pathways.nice.org.uk/pathways/hearing-loss

Hearing loss (adult onset)
In development [GID-QS10074]
Expected publication date: 10th July 2019
https://www.nice.org.uk/guidance/indevelopment/gid-q50074

Hearing loss prevalence in people who under-present for hearing loss: What is the prevalence of hearing loss among populations who under-present for possible hearing loss?
NICE: Individual research recommendation details
Recommendation ID: NG98/4

Please note that information provided in this update is collated from a variety of sources but coverage of the topic is not comprehensive.

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