Hearing in Patients with Intracanalicular Vestibular Schwannomas Per Caye-Thomasen, Thomas Dethloff, Søren Hansen, Sven-Eric Stangerup, Jens Thomsen Department of Oto-Rhino-Laryngology, Head and Neck Surgery, Gentofte University Hospital of Copenhagen, Copenhagen, Denmark

Abstract

This paper reports data on the spontaneous course of hearing in 156 patients with purely intracanalicular vestibular schwannomas. The mean pure tone average (PTA) was 51 dB HL and the mean speech discrimination score (SDS) 60% at diagnosis. The risk of a significant subsequent hearing loss (10 dB PTA or 10% SDS) was 54% during 4.6 years of observation. Patients with normal speech discrimination at diagnosis had a significantly smaller risk of losing hearing. The hearing loss at diagnosis and during observation was not related to age, gender, diagnostic tumor size, tumor-induced expansion of the internal auditory canal or tumor sublocalization (fundus, central or porus). However, the loss of PTA was smaller in shrinking tumors and the PTA deterioration rate correlated with the volumetric tumor growth rate. After 4.6 years observation, the PTA had increased by 14 dB to 65 dB HL, and the SDS reduced by 17% to 43%. The proportion of patients eligible for hearing preservation treatment as determined by word recognition score class I (70-100% SDS) was reduced to 28% (a 44% reduction), and by AAO-HNS class A to 9% (a 53% reduction).

Effect of Age and Cognition on Childhood Speech in Noise Perception Abilities Maria Talarico a, Geraldine Abdilla a, Martha Aliferis a, Irena Balazic a, Irene Giaprakis a, Toni Stefanakis a, Kate Foenander a, David B. Grayden b, Antonio G. Paolini a, b aSchool of Psychological Science, La Trobe University, Bundoora, and bThe Bionic Ear Institute, East Melbourne, Australia

Abstract

This research on children’s speech in noise and cognitive abilities aimed to determine the age-related trends in speech in noise perception abilities and the relationship between speech in noise perception and cognitive abilities. Monosyllabic distinguishable (consonant-vowel-consonant) words was the most recognisable word category, followed by monosyllabic confusable words (consonant-vowel-consonant), disyllabic non-words (/aCa/) and monosyllabic syllables (/Ca/), demonstrating that phoneme distinctiveness and a reduction in word confusability contribute to their recognition. Older children outperformed younger children on all speech in noise tasks, indicating that there are age-related trends in speech in noise abilities. Children with higher cognitive abilities did not outperform children with lower cognitive abilities on speech in noise tasks, indicating that the ability to hear speech in noise may be an intrinsic feature of the auditory system that matures with age.

Identification of Novel Mutations in the KCNQ4 Gene of Patients with Nonsyndromic Deafness from Taiwan Ching-Chyuan Sue d, Jiann-Jou Yang a,b, Jia-Ching Shieh b, Mao-Chang Sue c, Shuan-Yow Li e aGenetics Laboratory, Department of Biomedical Sciences, bInstitute of Medicine, Chung Shan Medical University, and cDepartment of Otorhinolaryngology, Head and Neck Surgery, Chung Shan Medical University Hospital, Taichung, and dTian-Sheng Memorial Hospital, Pin-Tong, Taiwan, ROC
Abstract

Ion channels play important roles in signal transduction and in the regulation of the ionic composition of intra- and extracellular fluids. Mutations in ion channels have long been thought to be responsible for some forms of hearing loss. Defects in KCNQ4, a voltage-gated potassium channel, are a cause of nonsyndromic sensorineural deafness type 2, an autosomal dominant form of progressive hearing loss. We present data of mutation analysis of KCNQ4 from 185 unrelated Taiwanese probands with nonsyndromic hearing loss. The analysis revealed three novel KCNQ4 mutations and many polymorphisms. The prevalence of KCNQ4 gene mutations in this study was 1.62% (3/185). The mutations include a missense mutation (F182L) and two silent mutations (R216R and T501T). The F182L missense mutation was located in the S3 domain of KCNQ4. The F182 residue of KCNQ4 is highly conserved in KCNQ4 among various species and is less conserved in all members of the KCNQ family. In addition, although R216R is a silent mutation and does not alter the content of amino acid residue, the neural network prediction system revealed that it can potentially create a novel splice donor site during transcription. This mutation might affect the protein structure of KCNQ4 and consequently the normal function of the K+ channel. Our data provide the first comprehensive analysis of the KCNQ4 gene in Taiwanese patients with nonsyndromic deafness.

Auditory Brainstem Implantation to Rehabilitate Profound Hearing Loss with Totally Ossified Cochleae Induced by Pneumococcal Meningitis

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Abstract

Hearing rehabilitation by cochlear implantation is not always possible in case of total ossification after pneumococcal meningitis. We report 3 cases of postmeningitis profound hearing loss with total cochlear ossification in adults who underwent auditory brainstem implantation (Nucleus 22®, Cochlear Inc., Lane Cove, Australia) between 1999 and 2004. The postoperative follow-up period ranged from 1 to 6 years. Eleven to 15 out of 22 electrodes were activated. All patients had significant speech discrimination in the sound-only mode and an enhanced lip-reading performance with the implant. Auditory brainstem implants are an efficient means of auditory rehabilitation and may be considered in selected cases of bilateral profound hearing loss with the impossibility of cochlear implantation.

Audiological Outcome of Infants with Congenital Cytomegalovirus Infection in a Prospective Study

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Abstract

The aim of this study was to evaluate the audiological outcome of long-term follow-up of infants with asymptomatic congenital cytomegalovirus (CMV) infection as defined by the presence of CMV DNA in neonatal urine. 12599 pregnant women underwent screening for CMV IgG and IgM antibodies between 1996 and 2003. Eighteen infants with congenital CMV infection were identified. These infants underwent the newborn hearing screening test or auditory brainstem response test. Follow-up hearing assessments were performed with the auditory brainstem response and behavioral audiometry. The seropositive rate of CMV IgG antibody among the pregnant women was 75.3%, and the yearly seropositive rate decreased over the study period. One hundred and forty-six pregnant women were positive for IgM antibody, and 18 neonates (12.3%) had congenital CMV infection. Sensorineural hearing loss (SNHL) was detected in 4 (25%) of the 16 infants with asymptomatic infection and 1 (50%) of the 2 infants with symptomatic infection during the first 6 months of life. Two infants who passed the newborn hearing screening had a delayed-onset SNHL in follow-up examinations up to 4 years of age. Two had progressive hearing loss and 2 had improvement of hearing loss. Screening of pregnant women for CMV infection and repeated audiological examinations of infants are necessary because there are infants with delayed-onset SNHL or improved SNHL caused by asymptomatic congenital CMV infection.

Cochlear Pharmacokinetics with Local Inner Ear Drug Delivery Using a Three-Dimensional Finite-Element Computer Model

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Abstract

Hypothesis: Cochlear fluid pharmacokinetics can be better represented by three-dimensional (3D) finite-element simulations of drug dispersal. Background: Local drug deliveries to the round window membrane are increasingly being used to treat inner ear disorders. Crucial to the development of safe therapies is knowledge of drug distribution in the inner ear with different delivery methods. Computer simulations allow application protocols and drug delivery systems to be evaluated, and may permit animal studies to be extrapolated to the larger cochlea of the human. Methods: A finite-element 3D model of the cochlea was constructed based on geometric dimensions of the guinea pig cochlea. Drug propagation along and between compartments was described by passive diffusion. To demonstrate the potential value of the model, methylprednisolone distribution in the cochlea was calculated for two clinically relevant application protocols using pharmacokinetic parameters derived from a prior one-dimensional (1D) model. In addition, a simplified geometry was used to compare results from 3D with 1D simulations. Results: For the simplified geometry, calculated concentration profiles with distance were in excellent agreement between the 1D and the 3D models. Different drug delivery strategies produce very different concentration time courses, peak concentrations and basal-apical concentration gradients of drug. In addition, 3D computations demonstrate the existence of substantial gradients across the scalae in the basal turn. Conclusion: The 3D model clearly shows the presence of drug gradients across the basal scalae of guinea pigs, demonstrating the necessity of a 3D approach to predict drug movements across and between scalae with larger cross-sectional areas, such as the human, with accuracy. This is the first model to incorporate the volume of the spiral
ligament and to calculate diffusion through this structure. Further development of the 3D model will have to incorporate a more accurate geometry of the entire inner ear and incorporate more of the specific processes that contribute to drug removal from the inner ear fluids. Appropriate computer models may assist in both drug and drug delivery system design and can thus accelerate the development of a rationale-based local drug delivery to the inner ear and its successful establishment in clinical practice.

**An Experimental Study of Tympanic Membrane and Manubrium Vibrations in Rats**

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**Abstract**

Rats are potentially very useful for auditory research because the middle ear structures are easily approachable and because rats are relatively inexpensive. The goal of the present study was to better characterize the mechanics of the rat middle ear by measuring frequency responses at multiple points on the tympanic membrane and manubrium. A laser Doppler vibrometer was used to measure the vibrations. Measurements were made on 7 rats. Tympanic membrane vibrations are presented for 7 different points in the frequency range of 1-10 kHz. The repeatability of the measurements and the interanimal variability at the umbo are also presented. The vibration modes of the tympanic membrane and manubrium were investigated.

**Vestibular Evoked Myogenic Potentials in Newborns**

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**Abstract**

This study presents a novel method for recording vestibular evoked myogenic potential (VEMP) in newborns, used to investigate the maturation of the sacculocollic reflex at birth. Twenty full-term newborns aged 2-5 days old were enrolled in this study. During natural sleep, each newborn underwent distortion product otoacoustic emission test, and VEMP test using the head rotation method. For comparison, 20 healthy adults also underwent VEMP test using the same method. Based on adult criteria, 40 newborn ears revealed normal VEMPs in 40%, prolonged VEMPs in 35%, and absent VEMPs in 25%, indicating that great variation exists in the maturation of the sacculocollic reflex at birth. Comparison of VEMP characteristics between healthy newborns and adults revealed nonsignificant difference in the response rate and the latency of n23. However, significant differences existed in the latency of p13, interpeak p13-n23 interval and p13-n23 amplitude between newborns and adults. In conclusion, VEMPs in newborns can be easily recorded by the head rotation method. Prolonged or absent VEMPs in newborns may reflect incomplete maturity of the sacculocollic reflex pathway, especially the myelination. A further large number of newborns receiving MRI scan for other reasons may undergo VEMP test to verify this hypothesis.