

Applying a Verbotonal Method to Rehabilitation of the Hearing Impaired Children

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P125**ACTIVIN SIGNALING DISRUPTION IN THE COCHLEA DOES NOT INFLUENCE HEARING IN ADULT MICE**Lukas Horvath¹, Daniel Bodmer², ²Vesna Radojevic, ¹Arianne Monge Naldi¹*Inner Ear Research, Clinic for Otolaryngology, Head & Neck Surgery, University Hospital Zurich, University of Zurich, Switzerland;* ²*Inner Ear Research, Clinic for Otolaryngology, University Hospital Basel, Switzerland*

Introduction. Activin, a member of the TGF- α superfamily, plays an important role in the development, repair and apoptosis of different tissues and organs and in the development of the cochlea. Activin binds to its receptor ActRII, then dimerizes with ActRI and induces a signaling pathway resulting in gene expression. A study reported a case of fibrodysplasia ossificans progressiva (FOP) with an unusual mutation in the ActRI gene leading to sensorineural hearing loss. This draws attention to the role of activin and its receptors in the developed cochlea. To date, only the expression of ActRII is known in the adult mammalian cochlea.

Methods. Transgenic mice with postnatal dominant negative ActRIB expression causing disruption of activin signaling *in vivo* were used for assessing hearing ability through auditory brainstem response (ABR) threshold and cochlear morphology.

Results. We present for the first time the presence of activin A and ActRIB in the adult mammalian cochlea.

Non-functioning ActRIB did not affect the ABR thresholds and did not alter the microscopic anatomy of the cochlea.

Conclusion. So we conclude that activin signaling is not necessary for hearing in adult mice under physiological conditions but may be important during and after damaging events in the inner ear.

P126**A NEW PATHOLOGICAL COCHLEAR PROFILE**

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Hearing loss associated with outer hair cell (OHC) degeneration is typically described by elevated hearing thresholds. Auditory function in mice is assessed by the combined use of auditory brainstem responses (ABR) and distortion product otoacoustic emissions (DPOAEs), with hearing threshold elevation relating to reduced or absent DPOAEs, evidence of OHC function loss. Moreover, in this pathological context, Masking Tuning Curves (MTC) show changes in shape included elevations or disappearance of the tip associated with the loss of frequency selectivity. In our study, we characterized the evolution of hearing profiles of two common laboratory mice (C57BL/6JRj and CD1 – RjOrl:SWISS) In this longitudinal study, we observed the progressivity of hearing impairment (at different rates in the two strains). However, a surprising discrepancy was found. ABR thresholds at high frequencies remained close to normal values but associated with absent DPOAEs. The MTCs show shifted tips centered on the low frequencies. These data indicate that basal OHC are no longer functional and the perception of high frequencies is disturbed. Histological observations with scanning electronic microscopy revealed abnormal stereocilia bundle at the cochlea base, with stereocilia persisting imprints on the lower face of the tectorial membrane. These data suggest cochleo-tonotopic disorganization.

P127**APPLYING A VERBOTONAL METHOD TO REHABILITATION OF THE HEARING IMPAIRED CHILDREN**Ivanka Jurjević- Grkinić, Boška Munivrana, Vesna Mijić
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The rehabilitation of listening in the principles of verbotonal method (VT) implies expansion of optimal hearing field of impaired ear. That means achieved hearing transfer i.e., transpose of listening from impaired frequency range to undamaged part. For example, ear which is strong impaired in the high frequency range obtain a capability for listening of words and sentences which belong to high frequency range. In the first case there is patient whose hearing impaired progressively because of many ear abscess (the main disease is Mucopolisaharidose tip II), this patient listens with hearing aids. While in the second case patient with the same history of the hearing impairment (progressively) at the age of nine received cochlear implant because of the strong hearing impairment. A scale was used as a test for the purpose of assessing listening of speech (EHS), (Jurjević, 2002), which is the result of many years of work with hearing impaired children, using the VT method and its basic principles about listening. The EHS scale is the easy and simple way of examining the listening of speech in the conditions of everyday spontaneous communication, in order to gain insight into the real possibility of listening of speech in all its components. The EHS scale examines listening of speech through ten levels from basic auditory recognition to precise listening and discernment of pairs of similar words. When choosing words, the frequency of speech material in the speech of children of certain chronological age was taken into account, as well as representation of congruent pitch words according to their appearance in the Croatian language. Tasks could be repeated up to three times, making sure that the repetition of a task does not undermine values of spoken language. The system of errors was analyzed as well as the number of repetitions in which the correct answer was reached. The results of this test give us the valuable data in further procedures of speech and hearing rehabilitation.

P128**HYPERACUSIS AND TINNITUS DEVELOPMENT OVER AGE AND THE INFLUENCE OF NOISE EXPOSURE IN THE RAT BEHAVIORAL MODEL**Sathish Kumar Singaravelu¹, Dorit Möhrle¹, Kun Ni², Dan Bing³, Ksenia Varakina¹, Mirjam Rieger¹, Marlies Knipper¹, Lukas Rüttiger¹¹*University of Tübingen, Department of Otolaryngology, Tübingen Hearing Research Centre (THRC), Molecular Physiology of Hearing, Tübingen, Germany;* ²*Shanghai Jiao Tong University, Department of Otolaryngology-Head & Neck Surgery, Shanghai Children's Hospital, Shanghai, China;* ³*Wuhan Jiefang Avenue No. 1095, Jong Ji Hospital, ENT Departemnt, Wuhan, China*

Aim. The prevalence of Tinnitus and Hyperacusis increases steadily due to changing leisure behavior and demographic trends. Tinnitus and Hyperacusis often occur after sudden hearing loss, e.g. through noise-induced hearing loss (NIHL), but also aging processes may lead to progressive hearing loss (age related hearing loss, ARHL). Previous studies demonstrated the degeneration of auditory fibers