group membership based on clinical symptoms. Comparable with this test, the headshake-impulse test has been put forward as a relevant simple test of the vestibular system at higher frequencies. It is supposed to have a better relationship with clinical symptoms than caloric tests and to be able to detect the side of vestibular pathology during clinical examination.

Non-syndromal autosomal dominant hearing loss


Sixteen forms of non-syndromal inherited hearing loss have been reported. Six of these can be distinguished clinically by means of type of audiogram, date of onset and/or severity of progression and have an autosomal dominant mode of inheritance. Recent investigations have revealed several gene localizations, for example on chromosomes 1p, 5q and 7p.

Two large unrelated families cooperated in a study to define hearing loss, to investigate vestibular function and to initiate gene-localization studies.

In family A (n = 179) 109 people were examined. Hearing loss could be attributed to an inherited cause in 44 persons. This number was 30 in family B (n = 129) in which 102 people were examined. No signs indicating an associated vestibular hypofunction were found. Linear regression analysis revealed an annual threshold increase of 1 dB in all frequencies with a slight interfamilial difference. Genetic studies were successful in both families and resulted in gene localization on chromosome 1p.

Analyses of audiologic studies of affected persons in one family can supply insight into the severity of progression of the hearing loss. The affected gene on chromosome 1p seems to play an important role in non-syndromal inherited hearing loss.

The bone-anchored hearing aid and the air conduction hearing aid compared


Increasingly, more patients who cannot use their air-conduction hearing aid any longer change over to the BAHA directly. The audiometric outcome of this transition is more difficult to predict. Until recently, audiometric results with the BAHA compared to the air conduction hearing aid were ambiguous. The small number of patients in a variety of studies prohibited conclusions.

In this study, the audiometric results of 34 patients fitted with a BAHA are presented. Patients’ performance with the individually fitted air-conduction hearing aid and BAHA was performed using audiometric tests and a questionnaire. The results of the speech recognition-in-noise test showed a significant improvement with the BAHA. This improvement was related to the size of the air–bone gap. The questionnaire demonstrated that the majority of the patients preferred the BAHA, most probably because fewer ear infections occurred. There was no evident preference in speech recognition.

When an air conduction hearing aid cannot be fitted, a BAHA is an alternative. Preoperative assessment of the size of the air–bone gap may be useful in predicting fitting results. It is important to inform the patient of the possibility of inferior subjective speech recognition.

Cisplatin ototoxicity and the possible protective effect of α-MSH in guinea pigs


Neuro- and ototoxicity are the dose-limiting side-effects of cisplatin (cis-diaminedichloro platinum II). Neurotoxicity can be delayed or prevented by simultaneous treatment with melanocortin-derived peptides like ORG 2766 and α-MSH. Recently, our group has found that also cisplatin ototoxicity can be reduced or prevented with ORG 2766. The present study was designed to investigate further the ameliorating effects of ORG 2766 on cisplatin ototoxicity and to evaluate the possibly similar effects of the physiologically more relevant parent peptide α-MSH.

Guinea pigs were injected (i.p.) with 2 mg/kg cisplatin for 8 consecutive days. α-MSH (75 mg/kg), ORG 2766 (75 mg/kg), or saline (controls) was given subcutaneously immediately before the cisplatin injection and an extra dose was given on day 9. Electrocochleography and hair cell counts were performed.

Cisplatin treatment and saline co-administration caused severe hearing loss (+60 dB at 8 kHz) combined with basal and medial turn hair cell loss in five out of six animals. However, in both the ORG 2766 and the α-MSH co-treated groups, three out of six animals could be classified as normal. We conclude that the effects of α-MSH and ORG 2766 co-treatment are comparable and that α-MSH might be clinically useful in protection against cisplatin-induced ototoxicity.

The perception of speech-in-noise by children

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Children with a conductive or sensorineural hearing loss have less intelligible speech. Some children with normal pure-tone