Vestibular function and motor skills in children with LVAS and IP2-malformation in relation to cochlear implantation
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Conclusions:
• Cochlear implantation is a safe method with low risk for postoperative vestibular disturbance
• Balance problems and vestibular disorders are common in patients with LVAS and/or IP2-malformation operated as children.
• A preoperative vestibular and balance test resulting in a prevention balance program for children at risk could be beneficial.
• This study stresses the importance of preoperative information on vestibular function and the risk for postoperative vertigo in children with LVAS and IP2-malformation.

Introduction
Vestibular function is affected to a higher extent in children with a sensorineural hearing loss and in particular in children with a malformed cochlea. Large Vestibular Aqueduct Syndrome, LVAS, with or without Incomplete Partition type 2, IP-2, is the most common and represents about 55% of the inner ear malformations.

Method
This cohort study includes 28 patients age 1.1-25.2 years with LVAS and/or IP2-malformation operated as children at a tertiary hospital setting. Pre- and postoperative testing with HIT, vHIT, minimal ice-water caloric test and c-VEMP was performed as well as a motor skill assessment with age-appropriate tests. Information regarding operation, type of implant and adverse events was collected from the charts.

Results
The compliance for vestibular testing was high (90%). 41 ears were operated, 5/26 (19%) ears had a vestibular loss preoperatively and 8/40 (20%) postoperatively. 1/10 (10%) operated ears with both pre- and postoperative testing lost vestibular function. 11/18 (61%) participants had reduced balance capacity at motor proficiency testing and in two patients with additional neuropsychiatric disorders it was severely affected. 12% had vertigo postoperatively and two of them had to stay another night at hospital due to extensive vertigo.

Discussion
This study show an high incidence of vestibular dysfunction and motor skill deficit in children with LVAS and/or IP2-malformation but a low risk for vestibular loss due to a cochlear implantation. Postoperative nausea and vertigo is a common adverse effect in this group and preoperative information is necessary. Vestibular testing can be performed from an early age and preoperative testing is important for preventive measures in form of preoperative information and balance training. Further research regarding the benefit of preventive balance program is needed.


Upcoming research from the authors:
• Vestibular function and method evaluation in newborns, Verrecchia et al
• Hearing, language and social skills in children with cochlear implants and LVAS and IP2, Bonnard et al
• Effect of preventive balance program in children with LVAS and IP2 malformation, Bonnard et al

V-HIT performed showing a pathologic vestibulo-ocular reflex (above) and a refixation saccade below in a 14 month old child.

MRI image of LVAS and IP2 malformation (left) and normal cochlea (right), LVAS marked with an arrow and the IP2 cochlea with its characteristic 1.5 turns.

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