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Vestibulo-cochlear symptoms due to large vestibular aquaeduct syndrome in children

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Large vestibular aquaeduct syndrome (LVAS) constitutes a rare but important nosological entity, responsible for balance disorders and/or cochlear symptoms in childhood.
The aetiology of this syndrome is related to deformity of the vestibular aquaeduct, acquired or genetic (syndromic or not). This occurs because the vestibular aquaeduct continues to develop after birth and reaches its final J-like shape at the age of 3 – 4 years. An enlarged vestibular aquaeduct, leads to propagation of intracranial pressure waves to the perilymphatic space, resulting to SN hearing loss and vertigo.
In other cases (e.g. X – linked anomaly with stapes gusher), the hearing loss seems to be of the mixed type.
Our material was based on five cases of LVAS, diagnosed the last five years. The patients' age varied between 4 and 18 years. Three of them were females and two were males. The localisation of the syndrome was bilateral.
In one of the cases, the hearing loss was moderate and in the other four cases it was modurate to profound.
In two of the cases, the vestibular disorder was manifested as recurrent vertigo attacks, in one as recurrent unsteadiness and in another as positional vertigo. One of the cases did not show any balance disorders.
In this study, we will present the audio-vestibular and laboratory findings and we will discuss the diagnostic procedure and the treatment possibilities.