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Case Report

A case of bilateral enlarged vestibular aqueducts and unilateral hearing loss at birth

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ABSTRACT

Screening of newborn hearing is nowadays widespread, and as many as 3.4% of all screened infants are found to have unilateral hearing loss. However, we, as clinicians and parents, usually dismiss the severity of unilateral hearing loss in pediatric patients. Recently, we experienced the case of unilateral hearing loss since birth in a 2-year-old girl who was finally found to have bilateral enlarged vestibular aqueducts when the disorder progressed to bilateral hearing loss. Proper evaluation of the cause of hearing loss, including imaging studies of the inner ear, and consecutive hearing evaluations should be mandatory in children with unilateral sensorineural hearing loss, due to the possibility of progression of hearing loss bilaterally.

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1. Introduction

Data from newborn hearing screening programs indicate that 0.5% to as many as 3.4% of all screened infants have unilateral HL. Considering possible underestimation of the actual prevalence in newborns owing to poor follow-up, relative amount of newborns assumed to have unilateral HL [1]. In children with unilateral sensorineural HL, the prevalence of inner-ear and/or internal auditory canal malformations detected by high resolution TBCT was reported very high from 25% to 67% [2–6]. Among their patients with anomalies, 50% had a corresponding anomaly of the unaffected ear. Around 10% of patients with unilateral sensorineural HL had a detriment in either the contralateral ear or both ears [3,5,7]. For this reason, all children with unilateral sensorineural hearing loss should have a TBCT scan, including genetic counseling and prognostic predictions of these cases. And regardless of whether a diagnosis is determined, the child should be followed closely with hearing test.

Enlarged vestibular aqueduct (EVA) syndrome is the most prevalent radiologically detectable inner ear anomaly in children with significant, permanent HL. Its HL is predominantly sensorineural HL of any degree. However, various degrees of mixed HL have also been observed [8]. The onset of HL in ears with EVAs is typically pre- or perilingual [9,10]. Most patients with EVAs have significant residual hearing, and in some cases this continues through to early childhood. The severity and progression of HL differs widely among individuals. However, it is not clear when HL due to EVAs begins or how much residual hearing remains at birth.

In this paper, we report the case of bilateral EVAs in a 2-year-old girl who showed unilateral sensorineural HL at birth and up to at least 6 months of age. We also review unilateral HL in pediatric patients and the hearing characteristics associated with EVAs.

2. Test procedures

At our institution, diagnostic hearing tests for infants include tympanograms, distortion-product otoacoustic emissions (DPOAEs), auditory brainstem response (ABR), and/or auditory steady state response (ASSR) evaluated in a sound booth (Industrial Acoustics Company, INC. Model 40A, Bronx, NY, USA). All infants are sedated with 50 mg/kg of Pocral® syrup (chloral hydrate, 100 mg/ml; Hanlim Pharm. Co., Ltd, Seoul, Korea) before testing.

Tympanograms are recorded with the GSI Tympstar version 2 (Grason-Stadler, Madison, WI, USA), and tympanometric data are obtained with a probe-tone frequency of 226 Hz. Ear canal pressure is swept in a positive-to-negative direction from +200 daPa to −400 daPa at a pump speed of 50 daPa/s. The following parameters are measured for each tympanogram: equivalent ear canal volume, tympanometric peak pressure, equivalent milliliters of compliance, and tympanometric width. The GSI Tympstar version 2 scaled automatically and presented data in equivalent ml of compliance at the peak compensated static acoustic admittance using 226-Hz tympanometry.

Distortion-product otoacoustic emissions are recorded using a GSI 60 Distortion Product Otoacoustic Emissions System (Grason-Stadler, Madison, WI, USA). The ratio of f_2/f_1 is fixed at 1.2

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throughout the experiments. The stimulus levels are also held constant at L_1 = 65 dB and L_2 = 55 dB SPL. The $2f_1 - f_2$ DPOAEs are depicted as a function of frequency in DPOAE-grams at three points per octave, that is, at eight f_1 frequencies ranging from 1000 to 5031 Hz. The DPOAEs are measured for each frequency and are considered to be present when they are at least 3 dB above the corresponding noise level.

Auditory brainstem responses are recorded with a GSI Audera (Grason-Stadler, Madison, WI, USA). Rarefaction clicks are presented to each ear through insert earphones at a rate of 33.1/s. The time window is set at 15 ms. Responses to air-conducted clicks are obtained at an intensity level of 70 dB nHL and the intensity is decreased by 20 dB nHL down to the level with no visible V Wave. At least two ABR waveforms are obtained at each level of intensity; responses to 500-Hz tone bursts and bone-conducted clicks are obtained with the infants appropriately sedated.

Auditory steady state responses are recorded with a GSI Audera (Grason-Stadler, Madison, WI, USA). The ASSR threshold is measured with an 81 Hz modulation frequency for each carrier frequency: 0.5, 1, 2, 4, and 8 kHz. Mixed modulation (100% amplitude modulation and 10% frequency modulation) is used. The ASSR thresholds determine the lowest intensity phase locked response at each carrier frequency, and the estimated audiogram window displays the predicted behavioral audiogram, which is extrapolated from the ASSR threshold data [11].

3. Case report

The subject was born at 37 weeks' gestation by Cesarean section; the birth was uneventful and she had a birth weight of 2.9 kg. Three days after her birth, a newborn hearing screening test was performed at the nursery using automated ABR. No issues were identified for her left ear, but she was referred for further evaluations of the right ear. A repeated automated ABR test performed 17 days after birth showed the same results. There was no family history associated with HL.

Diagnostic hearing evaluations with tympanometry, DPOAE, and ABR were performed at 40 days after birth. Middle ear measures indicated that the child had Type A tympanograms in both ears. Auditory brainstem responses to air-conducted clicks were obtained at decreasing intensities down to 70 dB nHL in the right ear and to 20 dB nHL in the left ear; DPOAEs were absent in the right ear and present in the left ear. A follow-up diagnostic hearing evaluation at the age of 6 months showed similar results; however, ABR to air-conducted clicks were obtained at 80 dB nHL in the right ear (Fig. 1). The girl was subsequently diagnosed with unilateral sensorineural HL on the right side and given a behind-the-ear hearing aid for that ear. Although we recommended that she be followed-up every 3 months, the child was not presented again until age 2 years when her mother became concerned with her delayed language development and unclear pronunciation.

Repeat diagnostic hearing evaluations at this time showed aggravated HL in both ears. Auditory brainstem responses to air-conducted clicks were obtained at decreasing intensities down to 90 dB nHL in the right ear and 50 dB nHL in the left ear. Responses to bone-conducted clicks were obtained at 50 dB nHL in the right ear and 10 dB nHL in the left ear. ASSR on the left side produced a descending type audiogram with high frequency hearing loss, which is unusual for conductive hearing loss (Fig. 2). Distortion-product otoacoustic emissions were absent in both ears, and Type A tympanograms were obtained for both ears. Temporal bone computed tomography (TBCT) revealed bilateral EVAs, but no other abnormalities of the inner ear (Fig. 3). We recommended bilateral hearing aids and aural rehabilitation. The girl's Infant

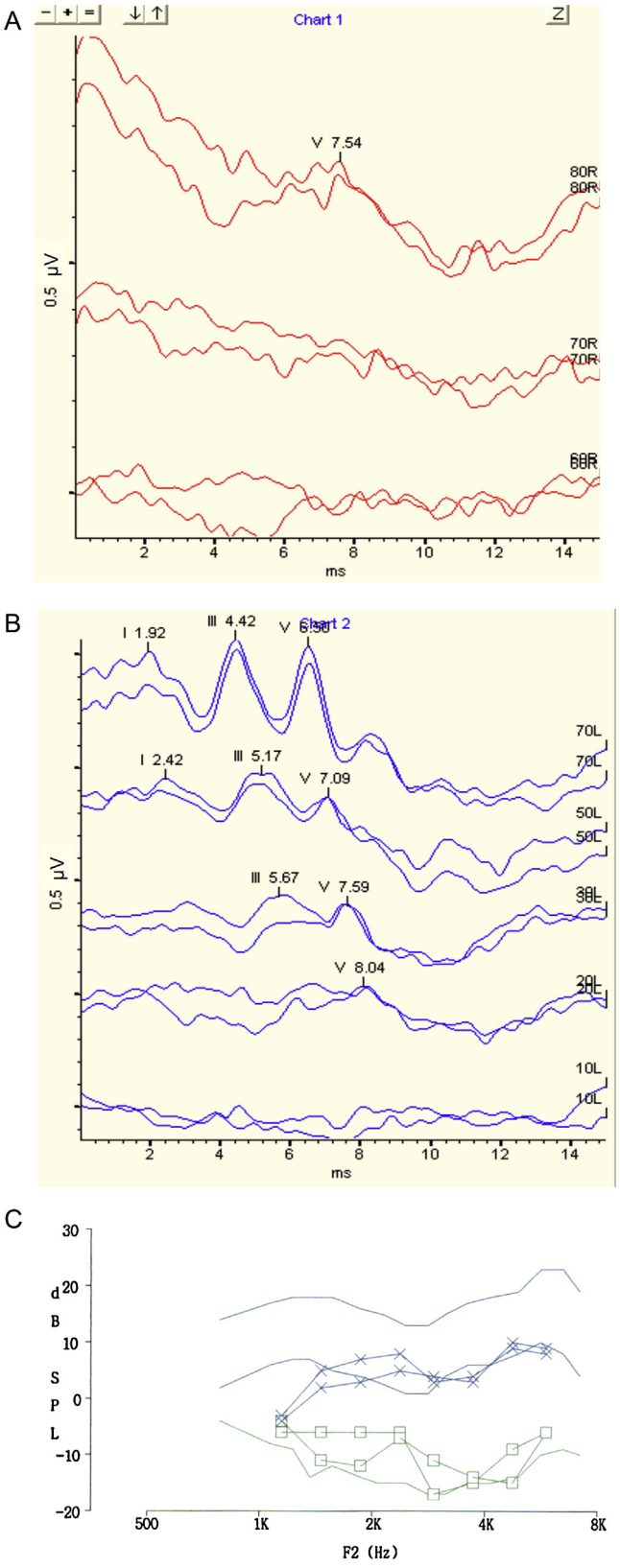

Fig. 1. ABR and DPOAE tests performed at 6 months of age. ABR to air-conducted clicks were obtained at decreasing intensities down to 80 dB nHL in the right ear (A) and 20 dB nHL in the left ear (B). DPOAE on the left side showed a good response (C).

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