International Journal of Pediatric Otorhinolaryngology 105 (2018) 16–19

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Pediatric Meniere's disease☆

Chi Wang[a], Chien-Hao Wu[b], Po-Wen Cheng[b], Yi-Ho Young[a,*]

[a] Department of Otolaryngology, National Taiwan University Hospital, Taipei, Taiwan
[b] Department of Otolaryngology, Far Eastern Memorial Hospital, Taipei, Taiwan

ARTICLE INFO

Keywords:
Cervical vestibular evoked myogenic potential (cVEMP)
Meniere's disease
Children
Ocular vestibular evoked myogenic potential (oVEMP)
Vestibular migraine

ABSTRACT

Objective: This study adopted an inner ear test battery comprising audiometry, and ocular vestibular evoked myogenic potential (oVEMP), cervical VEMP (cVEMP), and caloric tests to investigate the sequence of inner ear deficits in pediatric Meniere's disease (MD).

Methods: From 2005 to 2016, a total of 24 MD children aged < 15 years old underwent otoscopy, CT scan, blood test, and an inner ear test battery. Nine subjects were males and 15 were females, with mean age of 12 years. Right ear was affected in 3 patients, left ear in 5 patients, and both ears in 16 patients (67%). Eight (33%) of the 24 MD children had positive family history.

Results: Inner ear deficits in MD children ran from abnormal hearing (25%) to abnormal cVEMP (20%), oVEMP (13%), and caloric (12%) tests, exhibiting a significantly declining sequence from the cochlea, to the saccule, utricle and semicircular canals. Analysis between the laterality and family history revealed a significantly positive correlation.

Conclusion: Pediatric MD is rare, accounting for 2.3% prevalence in MD patients. The inner ear deficits in pediatric MD showed a significantly declining sequence from the cochlea, to the saccule, utricle and semicircular canals, mimicking the sequence in adult MD. One-third MD children had positive family history, which may explain the high rate of bilateral affliction and symmetrical hearing levels on both ears.

1. Introduction

Meniere's disease (MD) mostly affected middle-aged patients with a mean age of 40–50 years, and 10% of them had a disease onset after 65 years of age [1]. In contrast, MD rarely affected the pediatric age group, accounting for 1.5% of cases in a European report [2], 2.9% in an Asian report [3], and 3% in an American report [4]. Although the percentages of pediatric MD vary among continents, the consensus is that MD affects the pediatric group less frequently than adult group, as evidenced by sporadic case reports published in the literature during the last three decades [2–6]. The rarity of "definite" MD in pediatric patients is probably because small children cannot clearly describe their ear symptoms such as tinnitus and/or fullness sensation, which are essential for the diagnosis of definite MD. Additionally, aural symptoms in pediatric MD seem to develop over a long period of time, and diagnosis of MD in children is thus postponed. Alternatively, Brantberg et al. [5] suggested that young children with idiopathic recurrent vertiginous attacks more than 20 min, accompanied by fluctuating low-tone hearing loss may have definite MD.

An inner ear test battery comprising audiometry and caloric, ocular vestibular evoked myogenic potential (oVEMP) and cervical VEMP (cVEMP) tests provides insight into the localization and prevalence of hydrops formation in the inner ear compartments [7]. Huang et al. [8] reported that the decreasing order of percentages in abnormality of the cochlea, saccule, utricle and semicircular canals in adult MD mimicked the declining sequence of hydrops formation in temporal bone studies [9]. Since low-tone hearing loss and abnormal cVEMPs are identified in early stage of adult MD, whether pediatric MD manifesting as early stage of adult MD remains unclear. This study adopted an inner ear test battery comprising audiometry, and cVEMP, oVEMP, and caloric tests to investigate the sequence of inner ear deficits in MD children.

2. Patients and methods

From 2005 to 2016, a total of 1078 MD patients visited the neurotological clinic of the university hospital. Diagnosis of MD was based on the guidelines proposed by the American Academy of Otolaryngology-Head and Neck Surgery in 1995 [10], and new diagnostic criteria for MD defined by a multidisciplinary panel of experts from different disciplines and societies in 2015 [11]. However,

☆ This study was supported by National Science Council, Taipei, Taiwan (Grant no. MOST 105-2314-B002-163).
* Corresponding author. Department of Otolaryngology, National Taiwan University Hospital, 1 Chang-te St., Taipei, Taiwan.
E-mail address: youngyh@ntu.edu.tw (Y.-H. Young).

https://doi.org/10.1016/j.ijporl.2017.11.029
Received 9 September 2017; Received in revised form 27 November 2017; Accepted 29 November 2017
Available online 01 December 2017
0165-5876/

diagnosis of pediatric MD is modified from the literature [3,5], namely.

1) recurrent paroxysmal vertigo > 20 min or fluctuating cochlear symptoms
2) unknown etiology
3) no other neurological sign

Those with concurrent middle or inner ear anomaly/infection, previous ear surgery or head injury were excluded. Finally, 24 MD patients aged < 15 years old fulfilled the criteria and were enrolled, accounting for 2.3% prevalence in MD patients. Nine subjects were males and 15 were females, with their ages ranged 6-15 years (mean, 12 years). Right ear was affected in 3 patients, left ear in 5 patients and both ears in 16 patients. All patients received a detailed history taking followed by otoscopy, image study including CT of the temporal bone, blood test for syphilis, audiometry, and caloric test with electronystagmographic (ENG) recordings. After 2010, cVEMP and oVEMP tests were added to the test battery.

During the same period (2005–2016), totaling 374 pediatric patients (< 15 years old) with inner ear symptoms *i.e.* hearing loss or vertigo were experienced at the same clinic and were also included for comparison. Diagnoses of vestibular migraine (VM) and benign paroxysmal vertigo (BPV) of childhood were based on the guidelines proposed by the International Headache Society, 3rd edition and the literature [12,13].

This study was approved by the institutional review board, and each subject signed the informed consent to participate.

2.1. Audiometry

The hearing thresholds at the frequency of 125 through 8000 Hz were measured by an audiometer (Rion, AA67, Tokyo, Japan). Pure tone average (PTA) indicated averaged hearing threshold at four frequencies of 500, 1000, 2000 and 3000 Hz. In contrast, mean hearing level (MHL) was defined as mean hearing threshold at the same frequency from all MD ears.

2.2. Caloric test

The norm of slow phase velocity (SPV) of caloric nystagmus is 31 ± 12 (mean ± SD) °/s at our laboratory. Canal paresis is defined when mean SPV of caloric nystagmus in the lesion ear is < 7°/s, or as a greater than 25% difference between maximum SPV measurements for each ear, when compared with the sum of SPVs from each ear. If caloric response was not elicited, the subject underwent ice water (4 °C, 10 mL) caloric test to further confirm the caloric areflexia.

2.3. oVEMP test

Two active electrodes were placed around 1 cm below two lower eyelids. The other two reference electrodes were positioned 1-2 cm below the active ones, and one ground electrode was placed on the sternum. During recording (Smart EP 3.90, Intelligent Hearing Systems, Miami, USA), the subject was instructed to look upward at a small fixed target > 2 m from the eyes with a vertical visual angle of 30° above the horizontal. The stimulation rate was 5/s. The duration of each response was 50 ms, and 30 responses were averaged for each run.

BCV stimuli were delivered using a handheld electromechanical vibrator (minishaker 4810, Bruel and Kjaer, Naerum, Denmark). The input signal was 500 Hz sine wave, with initial peak driving voltage about 144 dB force level. The operator held the vibrator by hand and tapped on the forehead. If oVEMP was not elicited, alternatively, tapping at the ipsilateral mastoid site (2 cm behind the opening of external ear canal) was subsequently performed [14].

The initial biphasic waveform comprised peaks nI and pI. Consecutive runs were performed to confirm the reproducibility of peaks nI and pI, and oVEMPs were deemed to be present. The norm for the latency of peak nI was 11.4 ± 0.8 ms, and those with nI latency > 13.0 ms were defined as delayed response. The norm for the asymmetry ratio was 16 ± 12%, and those asymmetry ratio > 40% were interpreted as reduced response. In addition, those ears with absent oVEMP by forehead tapping but present oVEMPs by mastoid tapping were interpreted as reduced responses [14].

2.4. cVEMP test

Two active electrodes were placed on the sternocleidomastoid muscles; one reference electrode was on the suprasternal notch, and a ground electrode was on the forehead. The other settings were the same as in the oVEMP test except that the vibrator delivered a repeatable tap on the subject's head at inion [15,16]. The subjects elevated their heads during testing. To measure background muscle activity, subjects were required to keep a background muscle activity at 50-200 μV during data collection. The EMG signals were amplified and actual mean rectified background muscle activity for cVEMPs was measured. A total of 50 responses were averaged and recorded bilaterally.

The first positive and second negative polarities of biphasic waveform were termed waves p13 and n23, respectively. The norm for the latency of p13 was 14.4 ± 1.3 ms, and we defined when p13 latency > 17.0 ms as delayed cVEMP. The norm for the asymmetry ratio of cVEMP was 11 ± 11% and asymmetry ratio > 33% were interpreted as reduced response.

2.5. Statistical methods

The MHLs at each frequency were compared by one-way repeated measures ANOVA test, followed by Bonferroni-adjusted paired *t*-test. The abnormality rates among four tests were compared by Cochrane Q test. Analysis between laterality and family history was performed by McNemar test.

A p value < 0.05 indicates significant difference.

3. Results

From 2005 to 2016, we have experienced 374 pediatric patients with inner ear symptoms at the neurotological clinic. Most of them were vestibular migraine (VM) accounting for 188 patients (50%), followed by BPV of childhood in 115 patients (31%), MD in 24 patients (6%), sudden deafness in 15 patients (4%), and others in 32 patients (8%). Notably, 3 patients (1%) had posterior fossa tumor including arachnoid cyst in 2 and neuroblastoma in 1.

3.1. Clinical manifestation

Clinical manifestation in the 24 MD children comprised rotatory vertigo in 22 patients, and the remaining 2 patients had fluctuating cochlear symptoms. Other symptoms comprised nausea/vomiting (75%), tinnitus (62%), headache (46%) and fullness sensation (38%). Neither positive test for syphilis nor anomaly in CT scan was identified in all 24 MD children.

Eight (33%) of the 24 MD children had positive family history, while 16 patients had no family tendency. As regards to the laterality, 8 patients had one ear affected and 16 patients (67%) had both ears affected, totaling 40 MD affected ears. A significantly positive correlation was identified between the laterality and family history ($p = 0.0269$, McNemar test, Table 1).

3.2. Audiometry

Types of audiogram in the 24 MD children (40 MD affected ears) comprised low-tone hearing loss in 15 ears (38%), flat type in 13 ears (33%), down-sloping type in 8 ears, and saucer type in 4 ears.