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Review

Otolithic organ function in patients with profound sensorineural hearing loss

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Abstract

Profound sensorineural hearing loss (PSHL) is not uncommonly encountered in otology. In clinics, there is a high incidence of otolithic damage in patients with PSHL, but relevant reports are few. Sharing a continuous membranous structure and similar receptor cell ultrastructures, the cochlea and vestibule may be susceptible to the same harmful factors. Disorders of the inner ear may result in a variety of manifestations, including vertigo, spatial disorientation, blurred vision, impaired articulation, and hearing impairment. Considering the diversity of clinical symptoms associated with PSHL with otolithic dysfunction, it may be frequently misdiagnosed, and objective means of testing the function of otolithic organs should be recommended for hearing-impaired patients. Vestibular-evoked myogenic potentials (VEMPs) via air-conducted sound are of great importance for the diagnosis of otolithic function. Hearing devices such as cochlear implants are commonly accepted treatments for PSHL, and early identification and treatment of vestibular disorders may increase the success rate of cochlear implantation. Therefore, it is necessary to increase awareness of otolithic functional states in patients with PSHL.

Keywords: Profound sensorineural hearing loss; PSHL; Otolithic organs; Vestibular-evoked myogenic potential; VEMP

Contents

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1. Introduction

Profound sensorineural hearing loss (PSHL) is a specific form of sensorineural hearing loss (SNHL) caused by congenital or acquired lesions of the cochlea, auditory nerve and auditory center. The average air conduction audibility frequencies are 500, 1000, 2000and 4000 Hz, in which 0−25 dB represents normal hearing, 26−40 dB mild damage, 41−60 dB moderate loss; 61−80 dB severe damage, and more than 80 dB indicates profound hearing loss (Syka, 2002). PSHL is not uncommonly encountered in otology (Xu et al., 2016). The main features of PSHL are that it is most often associated with congenital hearing loss and infectious diseases. Congenital hearing loss is often bilateral and articulation is commonly impaired, as hearing loss often develops rapidly (Ozel et al., 2012). In addition to hearing loss, symptoms associated with the vestibular system may appear occasionally, such as nausea and vertigo. In children with PSHL, the incidence of otolithic damage is very high (Xu et al., 2015). It has been reported that the maximum possibility of discovering vestibular malfunction in children with hearing impairment is approximately 70% (Angeli, 2003), but this issue has not received sufficient attention from physicians. On account of the close physical connection between the cochlear and vestibular systems − for example, the cochlea and the saccule share the same membranous labyrinth (Sazgar et al., 2006) − they are frequently affected by the same factors. That said, the pathophysiology and causes of many conditions of the cochlear and vestibular systems remain to be fully determined.

Routine vestibular function tests such as Romberg's test, rotatory test and positional nystagmus test do not evaluate the function of otolithic organs. For this reason, it remains impossible to evaluate the degree of damage to the vestibular system definitively via these routine vestibular tests. In some situations, PSHL may potentially be accompanied by dysfunction of otolithic organs, which can be detected via vestibular-evoked myogenic potential (VEMP) responses (Xu et al., 2016). VEMP test, an electrophysiological examination, therefore deserves more attention. The main VEMP indexes include amplitude, latency and threshold. It can detect disorders of otolithic organs and the integrity of their pathways accurately, and is an easy and simple test of vestibular functions in clinical practice (Patko et al., 2003; Murofushi, 2016; Kim et al., 2015). PSHL with vestibular problems may affect individuals throughout their whole life, and may occur in infants as well as elderly individuals. Physicians should ensure that they are sufficiently knowledgeable in this area, in order that they may provide optimal treatment to this group of patients. Currently available hearing devices such as cochlear implants and hearing aids have facilitated great progress in hearing restoration. When physicians encounter PSHL, attention should be paid to otolithic functions during both diagnosis and treatment. VEMPs as a well-established test are particularly suited to the detection of latent otolithic dysfunction in patients.

2. Etiology and clinical manifestations in patients with PSHL

2.1. Etiology

Various causes of PSHL are encountered in otology clinics. The major risk factors for congenital PSHL include consanguinity, maternal rubella, and exposure to intrauterine infections, of which cytomegalovirus (CMV) infection is a significant cause of bilateral PSHL in children (Toumpas et al., 2014). Lack of an adequate vitamin A supply during pregnancy may result in the baby suffering developmental retardation in hearing (Emmett and West, 2014). Compared to congenital risks, the etiological factors of acquired PSHL are many and varied. Of them, bacterial meningitis is common among children (Karanja et al., 2013). A previously reported analysis of 310 adult cases included meningitis (24.4%), mumps (11.0%), unknown inflammatory diseases (16.6%), idiopathic sudden sensorineural hearing loss (ISSNHL; 19.4%), chronic suppurative otitis media (CSOM; 6.0%), trauma (6.1%), ototoxic medications (0.4%), and “no known cause” (16.1%) as causes of acquired PSHL (Ozel et al., 2012).

In addition to the above common factors, PSHL is often accompanied by dysfunction of vestibular organs (Wang et al., 2009). In one report, vestibular and cochlear symptoms occurred simultaneously in more than half of the patients (Gao et al., 2015). Given the similar physiological structures of the cochlear and vestibular organs, it is hypothesized that PSHL may prove to be significantly associated with vestibular disorders, especially in children (Cushing et al., 2008).

2.2. Clinical manifestations

In a recent clinical report on 29 patients with PSHL, neither the medical staff nor the patients themselves were mindful of vestibular dysfunction (Xu et al., 2016). Vestibular function can be normal or low in patients with PSHL. In one study, as the extent of hearing impairment increased, the detection rate reportedly gradually increased, although notably this result was not statistically significant (Gao et al., 2015). The cochlea and vestibule are closely related with regard to both anatomy and histoembryology, and both may be prone to the same risk factors. Notably, dysfunction of otolithic organs can cause a variety of serious problems including imbalance, dizziness, spatial disorientation and blurred vision. All these manifestations can be hidden and thus easily overlooked in patients with PSHL (Zhou et al., 2009).

Some patients with PSHL do not exhibit typical features of vestibular malfunction. For example, sometimes vertigo is not apparent even if the saccule is damaged (Hong et al., 2008). The most common chief complaint in PSHL patients may be “no response to sounds” (Xu et al., 2015), while vestibular dysfunction may also have a negative impact on the physical development in children with PSHL, in the form of delayed acquisition of head control or independent walking, or