

CASE REPORT

The “light cupula” phenomenon masquerading in congenital nystagmus

Kiyoshi Hiruma^{1*}, MD, Akihiro Ohara², MD, Izumi Koizuka², MD

Authors' affiliations:

1. Department of Otorhinolaryngology, Kawasaki Municipal Tama Hospital, Kawasaki, Japan
2. Department of Otorhinolaryngology, St. Marianna University School of Medicine, Kawasaki, Japan

* **Address for correspondence:** Kiyoshi Hiruma, MD, Dept. of Otorhinolaryngology, Kawasaki Municipal Tama Hospital, 1-30-37, Shukugawara, Tama, Kawasaki, 214-8525, Japan, Phone: +81-44-933-8111, Fax: +81-44-930-5181, E-mail: ZBN22654@nifty.com

Abstract

Objective/Hypothesis: Congenital nystagmus alone does not cause vertigo or dizziness, but we often encounter patients with this condition who complain of dizziness or vertigo. These patients also show positional nystagmus under an infrared camera (opened in the dark). Among them, we found a patient who had persistent direction changing positional nystagmus (DCPN) with two neutral points. This type of nystagmus is thought to be due to “light or heavy cupula” of the affected ear’s lateral semi-circular canal (SCC). The neutral point is deviated to the right or left from the supine position (first neutral point), and the deviated side is the affected side. If “light or heavy cupula” reflects the pathological condition of the endolymph in the affected ear lateral SCC, the side of the neutral point is thought to show the side of vestibular disorder. **Case Report:** The patient was a 51-year-old woman who complained of sudden onset of vertigo and hearing loss in the left ear. After admission therapy, hearing loss improved, but nystagmus did not disappear. She was diagnosed as having congenital nystagmus by typical electronystagmography. After this, she suffered vertiginous episodes and her left hearing level fluctuated and deteriorated. She was diagnosed with Menière's disease. Simultaneously, she also showed persistent DCPN with two neutral points (“light cupula” type was observed eight times and “heavy cupula” type was twice.), whose first neutral point’s side changed at each time. Finally, she continued showing “light cupula” type persistent DCPN with two neutral points. **Conclusion:** The clinical characteristics of our patient suggested that bilateral latent vestibular disorder can occur in patients with Menière's disease suffering from unilateral hearing impairment behind the congenital nystagmus. Persistent DCPN including “light cupula” can be explained by changes of the specific gravity of the surrounding endolymph in the affected ear.

Key Words: Congenital nystagmus, persistent_direction changing positional nystagmus (DCPN), neutral point, light cupula, Menière's disease

INTRODUCTION

Congenital nystagmus alone does not cause vertigo or dizziness, but we often encounter patients with this condition who complain of dizziness or vertigo. These patients also show positional nystagmus under an infrared camera (opened in the dark). Among them, we found a patient who had persistent direction changing positional nystagmus (DCPN) with two neutral points. This type of nystagmus is thought to be due to “light or heavy cupula” in the affected ear lateral semi-circular canal (SCC) [1-4]. The neutral point is deviated to the right or left from the supine position (first neutral point), and the deviated side corresponds to the affected side [1-4]. If “light or heavy cupula” reflects the pathological condition in the endolymph of the affected ear lateral SCC, the side of neutral point is thought to show the side of vestibular disorder. We report a patient who has suffered from Menière’s disease for a long time (1998.12~2010.9), and who showed “light or heavy cupula” type DCPN with the neutral point deviated to the affected ear at every episode of vertigo and hearing disturbance.

(Two types of persistent DCPN with neutral points; “light cupula type” and “heavy cupula” type)

At the neutral points, the cupula of the horizontal semicircular canal of the affected

ear is positioned vertical to the gravitational plane and no deflection of the cupula occurs. Additionally, in a position other than the neutral points, nystagmus beating away from the neutral point (in the geotropic direction) is thought to occur due to “light cupula”. On the other hand, in a position other than the neutral points, nystagmus beating towards the neutral point (in the apogeotropic direction) is thought to occur due to “heavy cupula”. The 1st neutral point is the position which deviates at an angle (20~30 degrees) from the supine position to the right or left side and the deviated side is thought to be the affected side. Thus, the 1st neutral point is considered to be pointing to the affected side [2-4].

Persistent geotropic DCPN (“light cupula” type) is different from transient geotropic DCPN (lateral SCC canalolithiasis). Lateral SCC canalolithiasis is transient with a latency of a few seconds, with fatigability, and shows no neutral point. On the contrary, “light cupula” is persistent without latency and fatigability.

CASE REPORT

The patient was a 51-year-old woman who complained of sudden onset of vertigo and hearing loss in the left ear. She was diagnosed with low-tone hearing loss (Fig. 1-A) and was admitted to a hospital where a steroid, prostaglandin E1, and diuretics were prescribed.

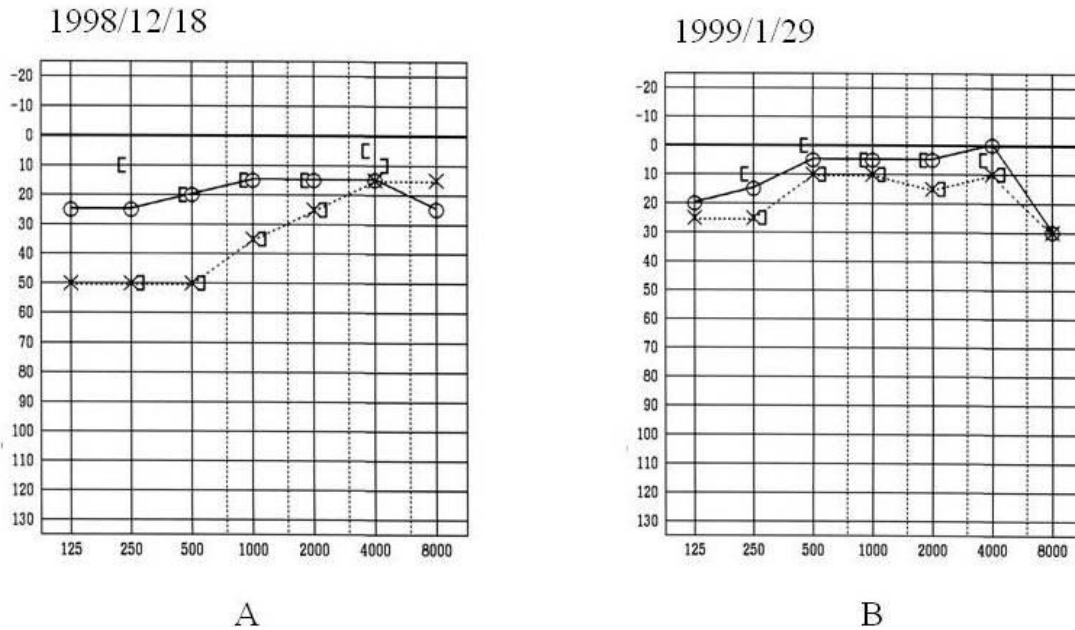


Figure 1-A. The patient was a 51-year-old woman who complained of sudden onset of vertigo and hearing loss in the left ear. She was diagnosed with low-tone hearing loss. **Figure 1-B.** She was prescribed a steroid, prostaglandin E1, and diuretics and hearing loss improved.



Fig 2

Figure 2. Her spontaneous nystagmus seemed to be pendular. Electronystamography (ENG) showed that this nystagmus disappeared when her eyes were closed in the dark while right beating nystagmus appeared during mental calculations. She was diagnosed as having congenital nystagmus.

Hearing loss improved (Fig 1-B), but nystagmus did not disappear, thus she was referred to our hospital. Her hearing level was normal at this time. Her spontaneous nystagmus seemed to be pendular. Electronystamography (ENG) showed that this nystagmus disappeared when her eyes were closed in the dark while right beating nystagmus appeared during mental calculations (Fig. 2) [5, 6]. Her optokinetic nystagmus pattern (OKP) test [7, 8] was inverted and jerking was noted superimposed on the eye tracking wave, suggesting congenital nystagmus (Fig. 3). This positional nystagmus was right beating. However, after it she experienced another

vertigo episode and her hearing level fluctuated and deteriorated (Figs. 4, 6). She was diagnosed with Menière's disease. Simultaneously, she showed persistent DCPN with two neutral points in which the first neutral point (1st NP) side changed (not only to the left side but also to the right side) at every attack (Figs. 4, 6) (right side was observed five times and left side was the same). Also, the type of persistent DCPN changed not only to "light cupula" type but also to "heavy cupula" type (Figs. 4, 6) ("light cupula" type was observed eight times and "heavy cupula" type was twice).

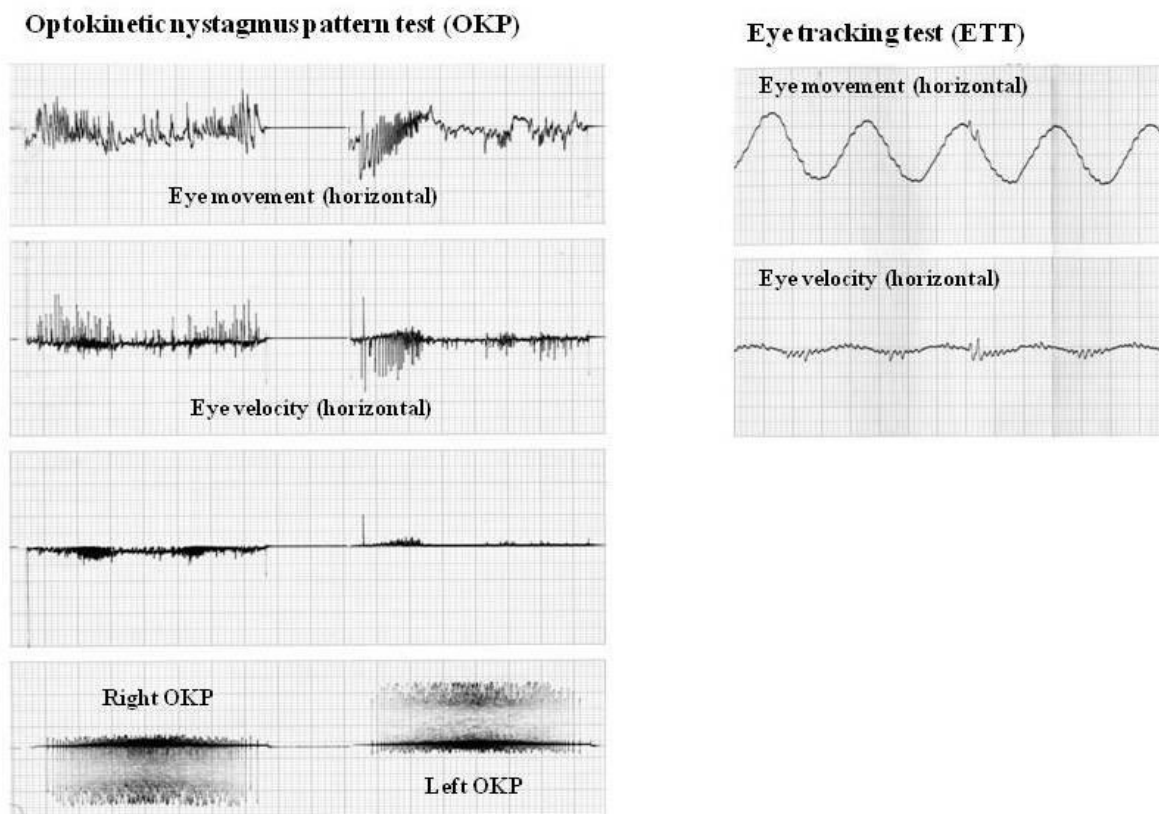


Figure 3. The results of an optokinetic nystagmus pattern (OKP) test suggested congenital nystagmus.

Figure 5 shows an ENG that was recorded with an infrared camera placed in goggles, while the patient had her eyes open in the dark to eliminate the effect of congenital nystagmus. At this time, ENG showed persistent geotropic DCPN (“light cupula” type) and the affected side was thought to be left side by 1st NP. We also performed a carolic test (iced water irrigation) during the ENG. The maximum slow phase velocity (MSPV) was 7 degrees/sec in the left ear

and 7.5 degrees/sec in the right ear. Therefore, we diagnosed her as having bilateral canal paresis (CP). About 12 years after she had visited us, her left hearing level was scaled out. Finally, the 1st NP moved to the left and she showed “light cupula” type persistent DCPN with neutral points (Fig. 6). She had no neurological disorders except for inner ear symptoms or central lesions revealed by magnetic resonance imaging (MRI) scan.

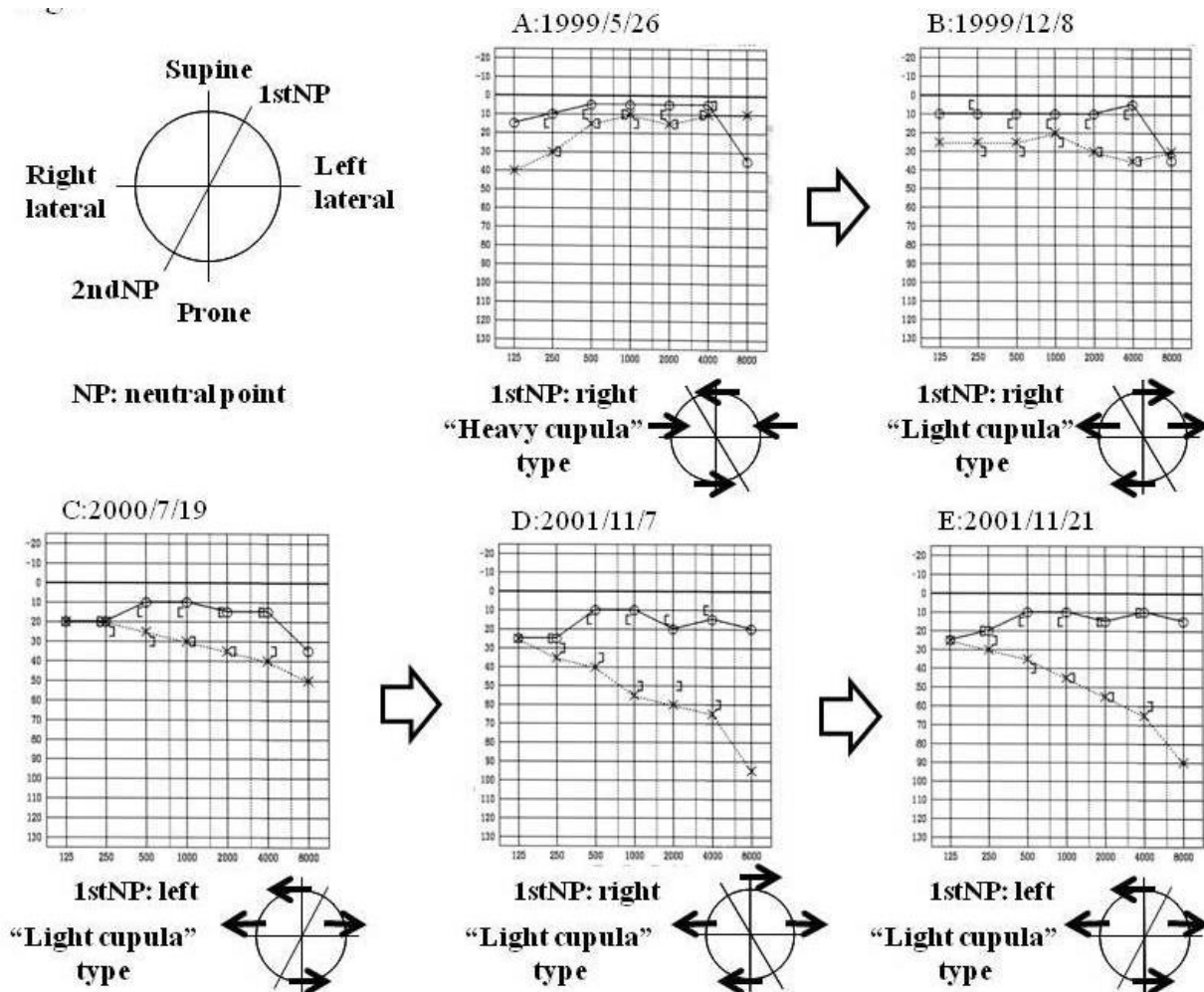


Figure 4. She experienced repeated vertiginous episodes and her left hearing level fluctuated and deteriorated. She was diagnosed with left Menière’s disease. Simultaneously, she also showed persistent DCPN with two neutral points in which the first neutral point side changed (not only to the left side but also to the right side) with every attack.

DISCUSSION

As congenital nystagmus disappeared when the eyes were closed or in the dark with loss of fixation [5], positional nystagmus recorded in the dark by an infrared camera placed in goggles is considered to be free from the effect of congenital nystagmus. Thus, this acquired positional nystagmus

would reflect a latent vestibular disorder. Especially, positional nystagmus beating to the same direction in every head position indicates imbalance between right ear and left ear. At times we observed right beating nystagmus (Fig. 2 and Fig. 6-C) in this patient, who showed a deterioration of the left vestibular function and was considered to be paralytic nystagmus.

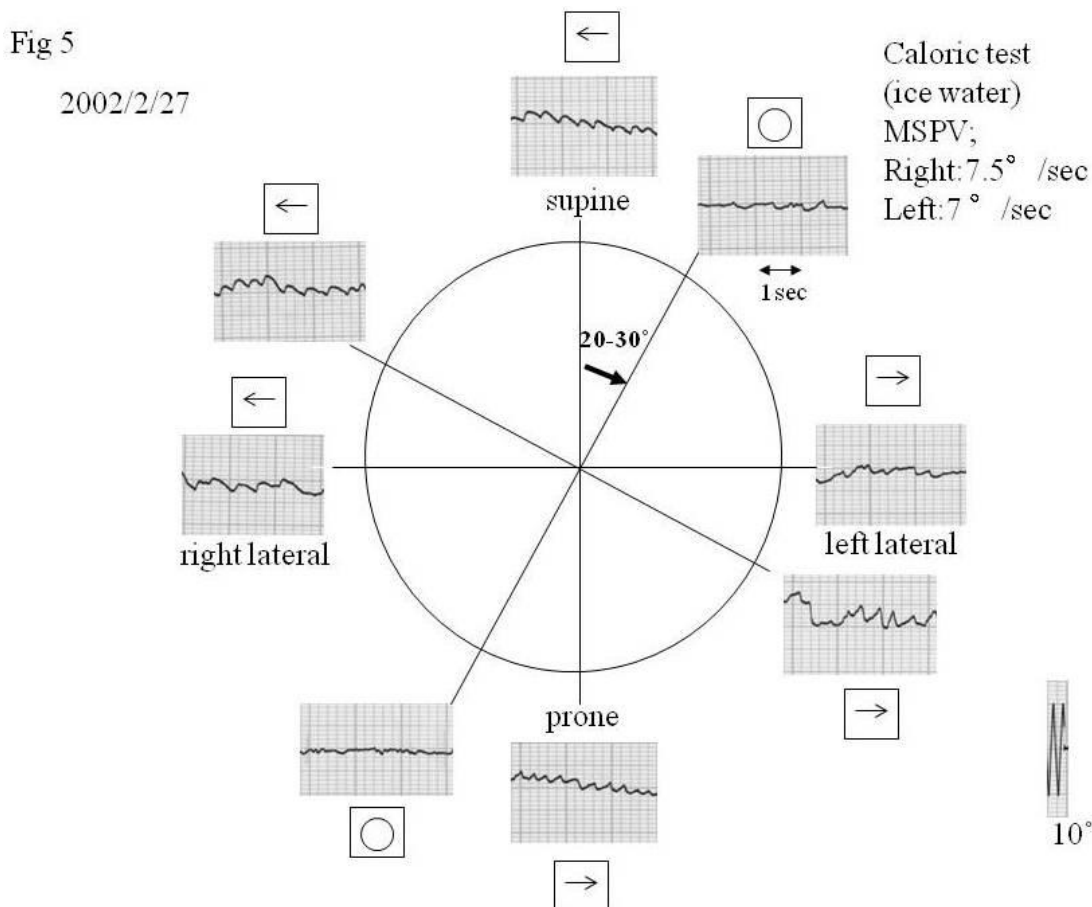


Figure 5. The positional nystagmus test revealed right-beating nystagmus in the supine position. When the head was rotated about 20-30° to the left from the supine position, nystagmus disappeared (the first neutral point). This positional nystagmus was canceled again when the head was rotated 180° from this position (the second neutral point). However, when the head was rotated through $\pm 90^\circ$ yaw from the neutral point, nystagmus became more intense. In addition, when the head was rotated to either side away from the first neutral point, nystagmus beating away from the first neutral point was observed (in the geotropic direction). This positional nystagmus was thought to be due to “light cupula”. We diagnosed bilateral canal paresis (CP) based on the results of the caloric test.

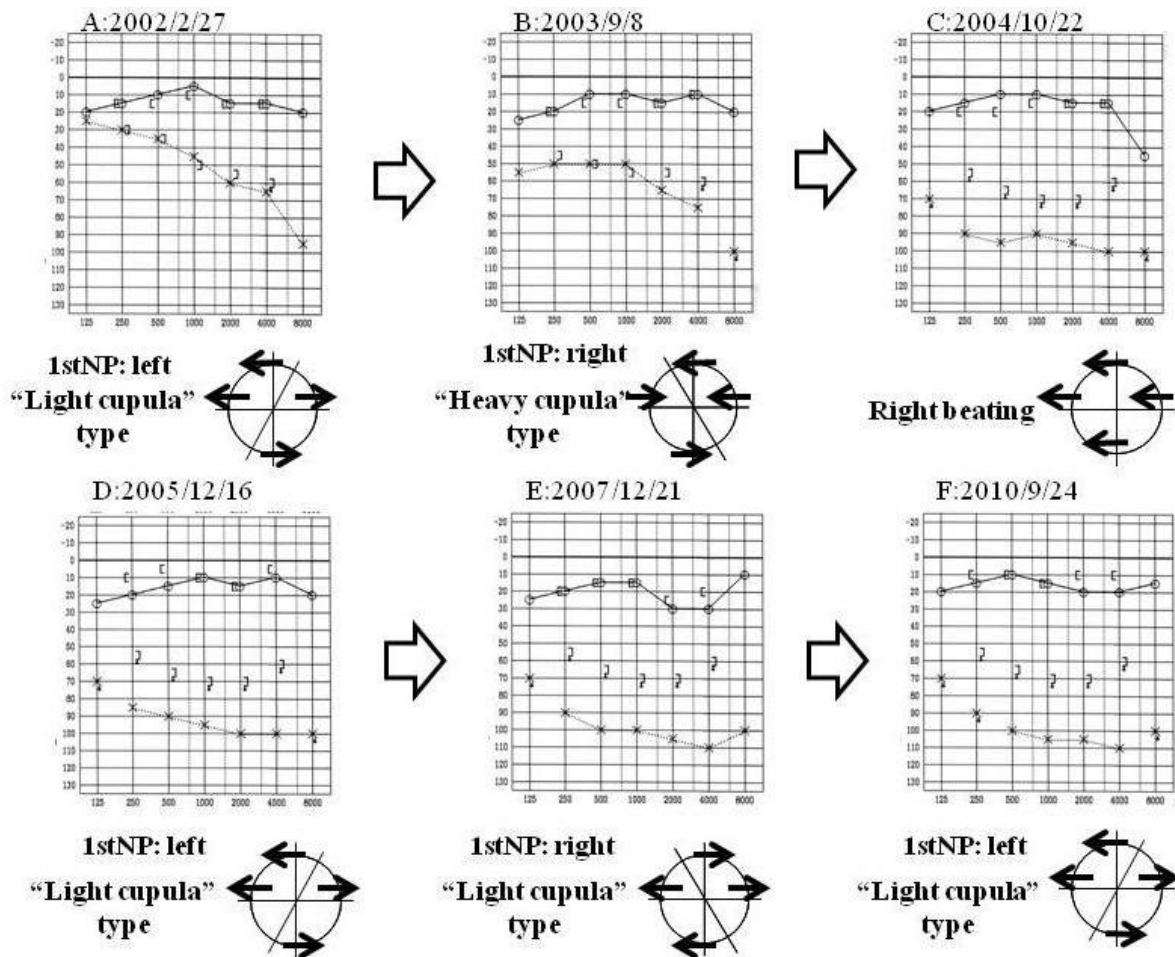


Figure 6. After continuing bilateral CP by caloric testing, the same DCPN persisted. The patient also experienced repeated vertiginous episodes and her left hearing level fluctuated and deteriorated. Simultaneously, she also showed persistent DCPN with two neutral points in which the first neutral point side changed (not only to the left side but also to the right side) with every attack. Finally, her left hearing level was scaled out and she showed "light cupula" type DCPN with two neutral points in which the first neutral point side was to the left.

Persistent DCPN with neutral points also emerged in our patient. She showed "heavy cupula" type or "light cupula" type. "Heavy cupula" type is thought to occur by the attachment of detached otoconia onto the cupula (cupulolithiasis), and "light cupula" type may be caused by the degree of buoyancy in the surrounding endolymph. But both "heavy cupula" and "light cupula" can be explained by changes of the specific

gravity of the surrounding endolymph in the affected ear [4, 9]. So some disorder of the endolymph besides endolymphatic hydrops may exist in this patient with Menière's disease. As "light cupula" type" (Fig.4-B, C, D, E, Fig.5, Fig.6-A, D, E, F), which cannot be explained by detached otoconia, occurred more frequently than "heavy cupula" type (Fig.4-A, Fig.6-B), we strongly believe an endolymph disorder

around the cupula was involved. Kim, et al. [4] speculated that blood plasma proteins leaking into the inner ear fluids due to breakage of the blood-labyrinthine barrier may increase the specific gravity of the endolymph. Additionally, the conversion between geotropic and apogeotropic persistent DCPN is assumed to be caused by an over-compensation of endolymphatic homeostasis [10]. On the contrary, Ichijo [3, 11] suggested “light debris”, which has not been identified as yet, are attached on the lateral SCC cupula and act as “light cupula” behavior. This hypothesis can also explain the conversion between geotropic and apogeotropic persistent DCPN.

About hearing impairment, Kim, et al. [12] reported the cases of sudden sensorineural hearing loss with simultaneous positional vertigo showing persistent geotropic DCPN. Because left side hearing was impaired in this patient, the 1st neutral point deviated to the left side. But it is hard to understand a deviation of the 1st neutral point to the right side, unless we assume that the right side was also affected. Consequently, in this patient the vestibular portion of the right ear was probably affected by some disorder while the cochlear portion of the right ear remained intact. Recently, it has become possible to diagnose bilateral Menière’s disease and recent studies have shown that unilateral disease can evolve toward bilateral Menière’s disease [13]. The

symptoms of our patient suggest bilateral Menière’s disease, occurring alternately at least in the vestibular portion. The neutral point is regarded as the indicator of the affected side.

After repeated vertigo and dizziness, this patient showed “heavy cupula” or “light cupula” type DCPN, right or left deviated neutral point at every attack, we performed a carolic test and confirmed bilateral canal paresis though not complete paresis. After this, she also showed “heavy cupula” or “light cupula” type DCPN, right or left deviated neutral point. Unless the canal works sufficiently, DCPN does not occur. And if superior vestibular nerve, from which lateral SCC derived, does not function completely, DCPN also would not occur. Thus, we thought that this phenomenon was caused by a morphological change of the cupula [14, 15] rather than by degeneration of the vestibular nerve itself. Viral infection, which is one of the causes of Menière’s disease, may have affected the function of the cupula. But the alternate occurrence on different occasions cannot be explained at all. Shin JE, et al. [16] reported the case of light cupula of the horizontal semicircular canal occurring alternately on both sides, but they said that pathophysiology is not entirely understood.

A pathological explanation of the behavior of the cupula (acting as “light cupula” or

“heavy cupula”) may be equivocal and controversial at present. Morphological and functional changes of the cupula may be a possible explanation. The relation between endolymph disorder around the cupula and endolymphatic hydrops is unclear. Because patients with Menière’s disease do not always show persistent DCPN with neutral points, these are considered different conditions. But a better understanding of the conditions of the endolymph around the cupula might contribute to solve this problem.

CONCLUSION

The present case suggested that bilateral latent vestibular disorder can occur in patients with Menière’s disease suffering from unilateral hearing impairment behind the congenital nystagmus. Persistent DCPN including “light cupula” can be explained by changes of the specific gravity of the surrounding endolymph in the affected ear, but the alternate occurrence on different occasions cannot be explained, especially in a patient with congenital nystagmus.

FINANCIAL DISCLOSURE

There is no financial support for this work. We do not have any financial interests in any companies or entities.

CONFLICT OF INTEREST

Conflict of Interest: None

References

1. Hiruma K, &Numata T. (2004). Positional nystagmus showing neutral points. *ORL J Otorhinolaryngol Relat Spec*, 66, 46-50.
2. Hiruma K, Numata T, Mitsunashi T, Tomemori T, Watanabe R, &Okamoto Y. (2011). Two types of direction-changing positional nystagmus with neutral points. *Auris Nasus Larynx*, 38, 46-51.
3. Ichijo H. (2012). Persistent direction-changing geotropic positional nystagmus. *Eur Arch Otorhinolaryngol*, 269: 747-751.
4. Kim CH, Kim MB, Ban JH. (2014). Persistent geotropic direction-changing positional nystagmus with a null plane: the light cupula. *Laryngoscope*, 124:E15-9.
5. Baloh RW, &Honrubia V. (1990).Electronystamography. Recording pathologic nystagmus; In: Baloh RW, Honrubia V (eds): *Clinical Neurophysiology of the Vestibular System*.2nd ed. Philadelphia: F.A.Davis Company, pp. 133-137.
6. Folstein MF, Folstein SE, &McHugh PR. (1975).“Mini-mental state” A practical method for grading the cognitive state of patients for the clinician. *J Psychiatr Res*, 12,

189-198.

7. Suzuki J, & Komatsuzaki A. (1962). Clinical application of optokinetic nystagmus. Optokinetic pattern test. *Acta Otolaryngol (Stockh.)*, 54, 49-55.
8. Yee RD, Baloh RW, & Honrubia V. (1980). Study of congenital nystagmus: optokinetic nystagmus. *Br J Ophthalmol*, 64, 926-932.
9. Casani A, Giovanni V, Bruno F, & Luigi GP. (1997). Positional vertigo and ageotropic bidirectional nystagmus. *Laryngoscope*, 107, 807-813.
10. Shin JE, Jeong KH, Ahn SH, & Kim CH. (2015). Conversion between geotropic and ageotropic persistent direction-changing positional nystagmus. *Acta Otolaryngol*, 135:1238-1244.
11. Ichijo H. (2016). Neutral position of persistent direction-changing positional nystagmus. *Eur Arch Otorhinolaryngol*, 273: 311-316.
12. Kim YW, Shin JE, Lee YS, & Kim CH. (2015). Persistent positional vertigo in a patient with sudden sensorineural hearing loss: a case report. *J Audiol Otol*, 19:104-107.
13. Nabi S, & Parnes LS. (2009). Bilateral Menière's disease. *Curr Opin Otolaryngol Head Neck Surg*, 17, 356-362.
14. Hiruma K. (2013). A case of an acoustic neuroma showing persistent geotropic positional nystagmus [in Japanese]. *Equilibrium Res*, 72: 17-21.
15. Seo T, Saito K, & Doi K. (2015). Intractable persistent direction-changing geotropic nystagmus improved by lateral semicircular canal plugging. *Case Rep Otolaryngol*, 2015, 192764.
16. Shin JE & Kim CH. (2015). Light cupula of the horizontal semicircular canal occurring alternately on both sides: a case report. *Ear, Nose & Throat Disorders*, 15:2.