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Pseudo-spontaneous nystagmus in horizontal semicircular canal canalolithiasis

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Abstract

Benign paroxysmal positional vertigo (BPPV) involving horizontal semicircular canal (HSCC) is characterized by direction-changing positional nystagmus (DCPN) in a supine roll test, and the occurrence of spontaneous nystagmus in HSCC BPPV has been reported recently. The aim of this study is to investigate the characteristics of pseudo-spontaneous nystagmus (PSN) in patients with HSCC canalolithiasis, and evaluate the effect of the presence of PSN on treatment outcome.

Between April 2014 and January 2016, 75 and 59 patients with HSCC canalolithiasis and cupulolithiasis, respectively, were enrolled. Spontaneous and positional nystagmus were examined.

PSN was observed in 31 of 75 patients (41%) with HSCC canalolithiasis, and 55 of 59 patients (93%) with HSCC cupulolithiasis. PSN persisted during the period of observation, which was at least 1 minute in all patients with PSN. In HSCC canalolithiasis, direction-reversing nystagmus was observed in 58 patients (25 bilateral and 33 unilateral). Nine of 25 patients with bilateral direction-reversing nystagmus, and 22 of 33 patients with unilateral direction-reversing nystagmus showed PSN. None of 17 patients without direction-reversing nystagmus showed PSN. The direction of PSN corresponded to that of direction-reversing nystagmus in all 22 patients with unilateral direction-reversing nystagmus. The proportion of patients who recovered after 1 session of repositioning maneuver was not significantly different between patients with and without PSN ($P = .867$).

PSN was observed more commonly in HSCC cupulolithiasis than canalolithiasis. The pathophysiologic mechanism underlying PSN can be explained by natural inclination of HSCC and medial to lateral orientation of the HSCC cupular axis in cupulolithiasis, and by spontaneous reversal of initial positional nystagmus (direction-reversing nystagmus) generated by short-term adaptation of vestibulo-ocular reflex in canalolithiasis. The presence of PSN in HSCC canalolithiasis may not affect the treatment outcome.

Abbreviations: BPPV = benign paroxysmal positional vertigo, CP = canal paresis, CRP = canalith repositioning procedure, DCPN = direction-changing positional nystagmus, HSCC = horizontal semicircular canal, PSCC = posterior semicircular canal, PSN = pseudo-spontaneous nystagmus.

Keywords: benign paroxysmal positional vertigo, direction-reversing nystagmus, horizontal semicircular canal, nystagmus, pseudospontaneous, short-term adaptation

1. Introductions

Benign paroxysmal positional vertigo (BPPV) involving the horizontal semicircular canal (HSCC) is characterized by direction-changing positional nystagmus (DCPN) in a supine roll test. When DCPN is directed toward the lowermost ear (geotropic) in a supine roll test, HSCC canalolithiasis can be diagnosed and gravity-dependent movement of otolithic particles within the HSCC is accepted as a possible mechanism. Geotropic DCPN starts after a few seconds' latency and lasts transiently. When the DCPN beats toward the uppermost ear (apogeotropic) during a supine roll test, otolith debris attached to the cupula is known to be a cause (HSCC cupulolithiasis). Apogeotropic DCPN is always persistent and lacks latency.

Recently, the occurrence of spontaneous nystagmus in HSCC BPPV has been reported, which showed widely diverse incidence.[1–13] Because spontaneous nystagmus is observed in the absence of unilateral vestibular impairment in many cases and the nystagmus disappears with resolution of BPPV, the term "pseudo-spontaneous nystagmus (PSN)" is commonly used to describe this nystagmus. While the occurrence of PSN in HSCC cupulolithiasis has been explained by cupular axis within the HSCC and influence of gravity,[3,13,14] the underlying mechanism of PSN in HSCC canalolithiasis is still under debate.

In the present study, we aimed to investigate the characteristics of PSN in patients with HSCC canalolithiasis and discuss short-term adaptation as its possible underlying mechanism. We also compared the treatment outcome between patients with and without PSN.

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2. Subjects and methods

2.1. Patients

We enrolled 75 consecutive patients (19 men and 56 women; aged 19–81 years) diagnosed with HSCC canalolithiasis and 59 consecutive patients (33 men and 26 women; aged 21–79 years) diagnosed with HSCC cupulolithiasis between April 2014 and January 2016. All patients reported repeated episodes of positional vertigo with changes in head position. Patients with otologic symptoms suggesting other labyrinthine diseases, and those with a recent history of labyrinthine disorders including sudden sensorineural hearing loss, vestibular neuritis, labyrinthitis, and Ménière's disease, or disorders of the central nervous system were excluded. We also excluded BPPV patients with multiple canal involvement and those with HSCC BPPV who exhibited both canalolithiasis and cupulolithiasis. The head impulse test revealed no catchup saccades in all patients, and neurologic examination did not detect any focal neurologic deficit in any patients. The barbecue roll maneuver was used as the canalith repositioning procedure (CRP) for treatment of the patients with HSCC canalolithiasis.[15] For patients with heavy cupula, the barbecue maneuver was performed after vibrating the mastoid bone on the affected side.

2.2. Evaluation of spontaneous and positional nystagmus

Spontaneous and positional nystagmus were evaluated and recorded using goggles installed with an infrared camera and a video-oculography system (EasyEyes, SLMED, Seoul, Korea or CHARTR VNG, ICS Medical, IL). Diagnosis of HSCC BPPV was made as follows: (1) in a seated position, spontaneous nystagmus was checked for at least 1 minute without change in head position, (2) in a seated position, the head was bent forward by 90° (bowing), and subsequently tilted backward by 60° (leaning) (bow and lean test), (3) in a supine position, the head was turned 90° to the right, returned to a neutral position, and then turned 90° to the left (supine roll test). The diagnosis of HSCC canalolithiasis was confirmed when patients showed typical transient geotropic nystagmus in a supine roll test, and the diagnosis of HSCC cupulolithiasis was confirmed when patients showed typical persistent apogeotropic nystagmus in a supine roll test.[16] Transient apogeotropic nystagmus in a supine roll test, which may indicate the presence of short arm HSCC canalolithiasis,[17] was not observed in any patient. Two experienced otolaryngologists blindly reassessed the eye movement recordings to confirm the presence of PSN. Direction-reversing nystagmus was determined to be present when the direction of initial positional nystagmus (geotropic nystagmus) spontaneously reversed (apogeotropic nystagmus) while the head position was maintained during a supine roll test in HSCC canalolithiasis.[10]

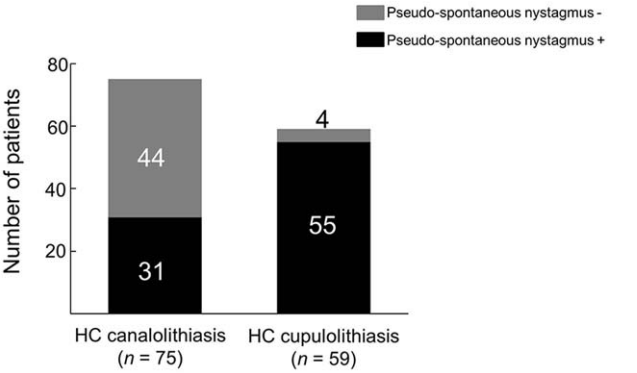

Figure 1. Incidence of pseudo-spontaneous nystagmus in patients with horizontal canal (HC) canalolithiasis (n=75) and cupulolithiasis (n=59).

The bithermal caloric test was performed in 12 of 31 HSCC canalolithiasis patients with PSN. The maximum slow-phase velocity of nystagmus was measured following irrigation, and Jongkees' formula was used to determine canal paresis (CP). A CP of 25% or more was considered abnormal, and, for all 12 patients, CP was less than 25%.

The study was approved by the Institutional Review Board (KUH1110049).

3. Results

PSN was observed in 31 patients (of 75, 41%) with HSCC canalolithiasis, and 55 patients (of 59, 93%) with HSCC cupulolithiasis (Fig. 1). PSN persisted during the period of observation which was at least 1 minute in all patients with PSN ($n=86$). In some patients with HSCC canalolithiasis, PSN was examined for longer than 3 minutes, and PSN persisted throughout the examination. In 55 cupulolithiasis with PSN, the nystagmus disappeared when the patient's head was bent forward about 30°. In 6 canalolithiasis patients with PSN, the patient's head was very slowly bent forward approximately 30° in an attempt to investigate if the nystagmus stops, and we could still observe PSN in all 6 patients.

In 55 patients with HSCC cupulolithiasis, PSN beat toward the side of null plane which corresponded to the affected side.[13,14] Among 31 patients with HSCC canalolithiasis, PSN was right-beating in 15 patients and left-beating in 16 patients (Table 1). Among 15 patients with right-beating PSN, bowing nystagmus was directed toward the left side in 6 patients, and leaning nystagmus was directed toward the right side in 6 patients

Table 1

Direction of PSN and positional nystagmus elicited by a supine roll test and a bow and lean test in HSCC canalolithiasis patients with PSN (n=31).

PSN ($n=31$)	Bow	Lean	Head-roll to right	Head-roll to left
RB (n=15)	RB (n=5)	RB (n=6)	RB with reversal (n=5)	LB with reversal (n=15)
	LB (n=6)	LB (n=4)	RB without reversal (n=10)	LB without reversal (n=0)
	No PN* (n=4)	No PN (n=5)		
LB (n=16)	RB (n=8)	RB (n=0)	RB with reversal (n=16)	LB with reversal (n=4)
	LB (n=0)	LB (n=8)	RB without reversal (n=0)	LB without reversal (n=12)
	No PN (n=8)	No PN (n=8)		

HSCC=horizontal semicircular canal, LB=left-beating, PN=positional nystagmus, PSN=pseudo-spontaneous nystagmus, RB=right-beating.
* "No PN" was defined when PSN was not changed by bowing or leaning maneuver.

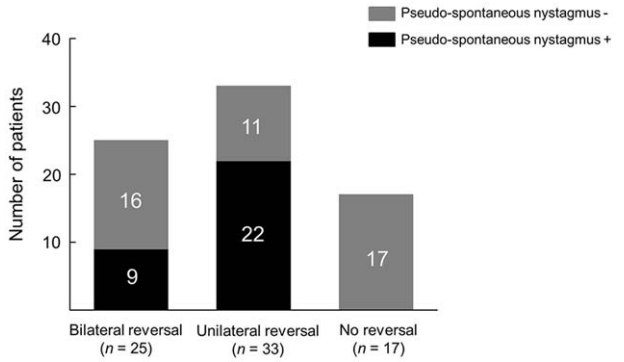

Figure 2. Incidence of pseudo-spontaneous nystagmus according to the presence of direction-reversing nystagmus during a supine roll test in patients with horizontal canal canalolithiasis (n = 75).

(Table 1). Among 16 patients with left-beating PSN, bowing nystagmus was directed toward the right side in 8 patients, and leaning nystagmus was directed toward the left side in 8 patients. Positional nystagmus was not elicited by bowing maneuver in 12 patients and by leaning maneuver in 13 patients among 31 HSCC canalolithiasis patients with PSN (Table 1).

Then, we investigated any relation between the direction of PSN and the presence of direction-reversing nystagmus during a supine roll test to analyze the direction of PSN in HSCC canalolithiasis. Among 75 patients with HSCC canalolithiasis, direction-reversing nystagmus was observed in 58 patients (58/75, 77%) of whom 25 showed direction-reversing nystagmus bilaterally and 33 showed it unilaterally (Fig. 2). Nine patients (of 25, 36%) with bilateral direction-reversing nystagmus showed PSN, and 22 patients (of 33, 67%) with unilateral direction-reversing nystagmus showed PSN. None of 17 patients without direction-reversing nystagmus showed PSN (Fig. 2). In cases with unilateral direction-reversing nystagmus, the reversal occurred on the side of stronger nystagmus intensity during a supine roll test,[10] and the direction of PSN corresponded to that of direction-reversing nystagmus in all 22 patients with PSN (Fig. 3). Among 9 patients with bilateral direction-reversing nystagmus, PSN beat toward the side of weaker and stronger nystagmus intensity during a supine roll test in 8 and 1 patients, respectively (Fig. 3). All 15 patients with right-beating PSN showed direction-reversing nystagmus in a left head-rolling, and all 16 patients with left-beating PSN showed direction-reversing nystagmus in a right head-rolling (Table 1). Direction-reversing nystagmus was observed in a bowing and/or leaning maneuver in some patients.

Treatment outcome was compared between HSCC canalolithiasis patients with and without PSN. Twenty-two patients (of 31, 71%) with PSN and 32 patients (of 44, 73%) without PSN recovered after 1 session of repositioning maneuver (Table 2), and the proportion of patients who recovered after 1 session of repositioning maneuver was not significantly different between patients with and without PSN ($P = .867$, Chi-square test; Fig. 4).

Table 2

Treatment outcomes of the patients with horizontal canal canalolithiasis (n = 75).

	1 session	2 sessions	3 or more sessions
PSN (+) (n = 31)	22	8	1
PSN (−) (n = 44)	32	7	5

PSN = pseudo-spontaneous nystagmus.

4. Discussion

In this study, we examined 75 patients with HSCC canalolithiasis and 59 patients with HSCC cupulolithiasis, and found that 41% (31/75) of HSCC canalolithiasis and 93% (55 of 59) of HSCC cupulolithiasis showed PSN. Previously reported incidence of PSN in HSCC BPPV has been widely variable. The presence of PSN was reported in 64% to 76% of patients with HSCC BPPV from the studies that included both geotropic and apogeotropic type.[1,6] PSN was observed in 14% to 67% of HSCC canalolithiasis and 16% to 71% of HSCC cupulolithiasis.[5,7–9] The incidence of PSN in our patients with HSCC cupulolithiasis was higher than that of previous reports. Considering that the anterior part of HSCC is tilted upwards approximately 30° from the horizontal plane and the axis of HSCC cupula is oriented in a medial to lateral direction,[3,5,13,14] the HSCC cupula would be deflected towards the utricle in an upright-seated position, which

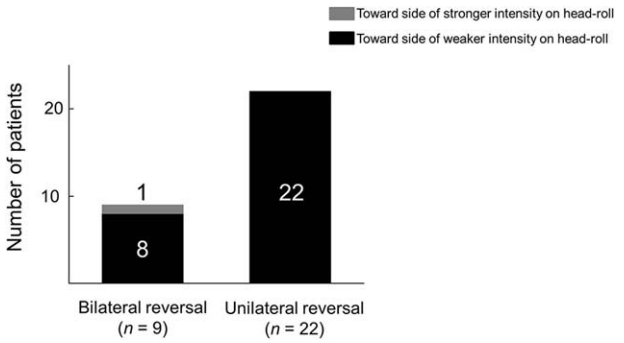

Figure 3. The direction of pseudo-spontaneous nystagmus in patients with horizontal canal canalolithiasis showing direction-reversing nystagmus bilaterally (n = 9) or unilaterally (n = 22).

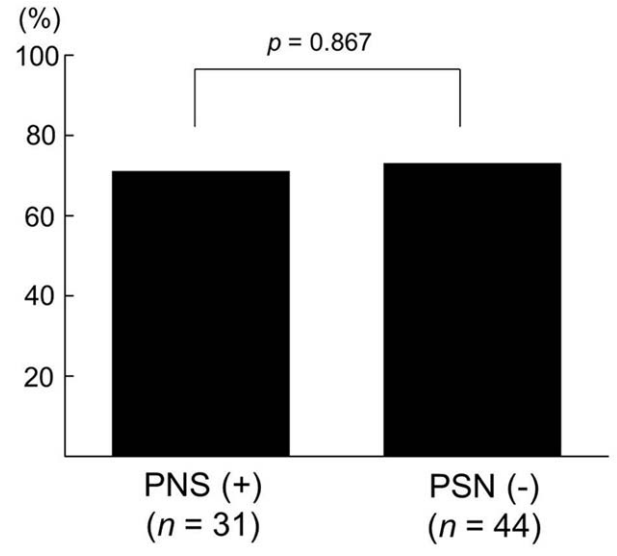

Figure 4. Comparison of treatment outcomes between patients with horizontal canal canalolithiasis showing pseudo-spontaneous nystagmus and those not showing pseudo-spontaneous nystagmus.

may reasonably explain the high incidence of PSN beating toward the affected side in HSCC cupulolithiasis.

It has been proposed that PSN in HSCC canalolithiasis is caused by slow movement of otolith particles within the HSCC under influence of gravity in a upright-seated position because the anterior part of HSCC is upwardly inclined approximately 30° from the horizontal plane.[1,6–9,18] In some studies, the direction of PSN in HSCC canalolithiasis was contralesional in all patients,[6,8] while in others, it was either contralesional or ipsilesional.[1,7,9] It was described that when HSCC canalolithiasis is geotropic form (otoliths settled in posterior arm of the HSCC) PSN beats toward the contralesional side, and when HSCC canalolithiasis is apogeotropic (otoliths settled in anterior arm of the HSCC) PSN beats toward the ipsilesional side.[1,9,18] When the patient's head was bent forward about 30° to make the HSCC parallel to the horizontal plane, PSN disappeared due to cessation of otolith movement within the HSCC.[1,6] Regardless of whether HSCC canalolithiasis is geotropic type or apogeotropic type, the direction of PSN would theoretically be identical to that of bowing or lying-down nystagmus, which rendered it to be proposed as one of the clinical signs for determining the affected side,[1] even though the frequency of PSN in HSCC canalolithiasis has been reported to vary from 14% to 67%.[5,7–9] To detect PSN with increased accuracy at the outpatient clinic, it was suggested that sufficient amount of time should be devoted to examining PSN, and patient's head position should be precisely controlled.[8] Other distinguishing characteristics of PSN are that it is a particularly long lasting, and mild head shaking may increase the occurrence of PSN.[1,18] However, some findings cannot be clearly explained solely by this hypothesis which is based on natural inclination of the HSCC and otolith movement; (1) the frequency of PSN beating toward the ipsilesional side in HSCC canalolithiasis was higher than expected,[7,9] (2) the direction of PSN was not in accord with that of lying-down nystagmus in some patients,[8] (3) PSN did not disappear in a relatively short time.[1,18]

In the present study, we considered the possibility of relation between PSN and direction-reversing nystagmus in patients with HSCC canalolithiasis based on the findings that PSN was observed only in patients who showed direction-reversing nystagmus during a supine roll test (Fig. 2). In addition, PSN exhibited prolonged duration and was persisted until the end of the examination, as observed in direction-reversing nystagmus.[10,19] Direction-reversing nystagmus, which is thought to be generated by short-term adaptation of vestibulo-ocular reflex, was observed in either bilateral ($n=25$) or unilateral ($n=33$) head-rolling, and in unilateral cases, reversal occurred on head-rolling to the side of stronger initial nystagmus intensity.[10,20] The direction of PSN was identical to that of direction-reversing nystagmus in all 22 patients who showed PSN and unilateral direction-reversing nystagmus, and was identical to that of direction-reversing nystagmus elicited by head-rolling to the side of stronger initial nystagmus intensity ($n=8$) or by head-rolling to the side of weaker initial nystagmus intensity ($n=1$) in patients with bilateral direction-reversing nystagmus (Fig. 3). It can be speculated that PSN is observed when the prolonged direction-reversing nystagmus is elicited by recent stimulation or inhibition of the affected HSCC, and thus the direction of PSN represents that of prolonged direction-reversing nystagmus. Therefore, in patients with bilateral direction-reversing nystagmus, the direction of PSN can be either side,[7,9] and PSN beating towards the side of weaker intensity on head-rolling can be more easily detected[6,8] as more robust direction-changing nystagmus is observed on the side of stronger initial nystagmus intensity.[10] Furthermore, previous findings that PSN was not observed in patients with posterior semicircular canal (PSCC) BPPV[6] might be explained by the observations that direction-reversing nystagmus rarely occurred in cases with PSCC BPPV.[10] It has been suggested that PSN can provide helpful information for lateralization of HSCC canalolithiasis because, in theory, the direction of PSN corresponds to that of leaning or lying-down nystagmus.[1,5] However, the present study demonstrated that only 14 out of 31 patients with HSCC canalolithiasis had PSN and leaning nystagmus beating toward the same direction (Table 1) as reported in the previous study.[8] Moreover, because not only PSN but also leaning or lying-down nystagmus is not observed in all patients with HSCC canalolithiasis,[10,21,22] comparative interpretation of the results of a supine roll test, bow and lean test, and PSN would be essential in determining the affected side in HSCC canalolithiasis.[10,11] In our study, a caloric test was performed in 12 patients with PSN, which revealed no canal paresis in all patients. The fact that only 39% (12 of 31) of patients with PSN took a caloric test may impose limitation on the validity of this study because caloric weakness might have been superimposed on HSCC BPPV causing spontaneous nystagmus, even though clinical tests including head impulse test revealed no signs suggesting unilateral vestibulopathy. The present study demonstrated that treatment outcome between patients with PSN and those without was not significantly different, which was consistent[6] or inconsistent with the previous studies.[8,9]

5. Conclusion

PSN was observed more commonly in HSCC cupulolithiasis (55 of 59, 93%) than HSCC canalolithiasis (31 of 75, 41%). Pathophysiologic mechanism underlying PSN can be explained by natural inclination of HSCC and medial to lateral orientation of the HSCC cupular axis in cases with cupulolithiasis, and can be explained by spontaneous reversal of initial positional nystagmus (direction-reversing nystagmus) generated by short-term adaptation of vestibulo-ocular reflex in cases with canalolithiasis. The presence of PSN in HSCC canalolithiasis may not affect the treatment outcome, but future study with a large number of patients would be required to clarify the prognostic importance of PSN.

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