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Prognostic Value of Initial Electronystagmography Findings in Idiopathic Sudden Sensorineural Hearing Loss Without Vertigo

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Objective: We used electronystagmography (ENG) to characterize recurrent hearing loss and its progression to definite Ménière's disease in patients with idiopathic sudden sensorineural hearing loss (SSHL) without subjective vertigo.
Methods: We reviewed medical records of 1,334 patients with unilateral hearing loss initially diagnosed with idiopathic SSHL between 1985 and 2003 at our university hospital. Of the 1,334 patients, we examined 127 (86 with low-tone and 41 with high-tone SSHL) who underwent ENG within 30 days of the initial diagnosis and who could be followed up during the long term.
Results: Spontaneous nystagmus (SN) was observed in approximately half of the vertigo-unaccompanied group. Long-term follow-up with a mean of 67 months revealed that the recurrence rate of hearing loss was 51.2% in low-tone SSHL patients with SN. The recurrence rate of hearing loss was 27.9% in low-tone SSHL patients without SN. Progression to Ménière's disease occurred in 14.0% of the low-tone–type and 12.5% of the high-tone–type patients when SN was detected. In contrast, in both the low-tone–type and high-tone–type groups, none developed Ménière's disease when SN was absent.
Conclusion: Our results suggest that the initial ENG findings could provide prognostic information for idiopathic SSHL patients, even those who have no vestibular symptoms at the first visit. **Key Words:** Endolymphatic hydrops—Low-tone hearing loss—Recurrent hearing loss—Spontaneous nystagmus—Vertigo.

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Sudden sensorineural hearing loss (SSHL) has a variety of causes, including viral infection, ototoxic substances, neoplasms, head injury, autoimmune disease, and vascular disorders. When the underlying cause is unidentified, SSHL may be clinically diagnosed as idiopathic. However, SSHL of low-tone type with preservation of high-tone hearing and no episodes of vertigo may be distinct from idiopathic SSHL (1–5). In 1982, Abe (1) first proposed the clinical entity of acute low-tone sensorineural hearing loss (ALHL), characterized as deterioration in audiometric low frequencies (125, 250, and 500 Hz), preservation of high frequencies (2, 4, and 8 kHz), and no episode of dizziness and/or vertigo (2). More than 80% of ALHL patients show good hearing recovery within 3 months (3,4). However, 27% of long-term ALHL patients have subsequent recurrent hearing, and 11% of these patients progress to Ménière's disease (4). Electrocochleography (ECochG) and glycerol tests suggest that the pathophysiology of ALHL is attributable to endolymphatic hydrops confined to the cochlea (4–6).

We reviewed 250 SSHL patients retrospectively with a long-term follow-up. We found that ECochG at the initial examination can aid in determining the prognosis for recurrent hearing loss (7). The average ratio of the summating potential/action potential of the ECochG was larger for SSHL patients with recurrent episodes than for those with a single episode. No statistically significant difference was evident in the average summating potential/action potential between low-tone and high-tone SSHL (7). This result suggests that cases of ALHL with recurrent episodes might have endolymphatic hydrops. Cases characterized by only a single episode are more likely attributable to some other cause.

Several factors such as the initial audiogram pattern, hearing level, age, and the presence or absence of complications such as diabetes and cardiovascular disease may affect hearing recovery from idiopathic SSHL (8–13). The presence of vertigo and spontaneous nystagmus (SN) correlate with recovery from SSHL (8,14). However, there have been few studies demonstrating the value of initial vestibular testing for predicting the

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long-term prognosis of SSHL patients, especially for those with no vertigo sensation with/without SN.

The purpose of the present study was to investigate whether or not a simple vestibular examination can be useful for the prognosis of low-tone SSHL patients, especially in those who do not complain of vertigo. Consequently, we first investigated the rate of vestibular alterations in SSHL patients. Subsequently, we compared the recurrence rate of hearing loss and the progression rate to definite Ménière's disease in SSHL patients with and without SN. Our results indicate that SN recorded by electronystagmography (ENG) predicts progression to Ménière's disease.

MATERIALS AND METHODS

Medical records of 1,334 patients with unilateral hearing loss who were initially diagnosed with idiopathic SSHL between 1985 and 2003 were reviewed retrospectively. We defined idiopathic SSHL as hearing loss for which the sum of the hearing levels at 3 consecutive audiometric frequencies was 70 dB or more, developed within 24 hours or less, and was not attributable to any commonly identifiable event.

Of the 1,334 patients, idiopathic SSHL patients who met all of the following inclusion criteria were selected: 1) no previous hearing loss; 2) no history of ocular or vestibular disease; 3) ENG performed within 30 days of the initial diagnosis; 4) medical treatment received within 30 days of the onset; and 5) patient availability for long-term follow-up.

Several exclusion criteria were also applied. Patients with central and retrocochlear hearing loss as measured by auditory brainstem response and magnetic resonance imaging were excluded. As part of our battery of clinical neurootologic examinations, computer analysis of ENG recordings was routinely performed for all idiopathic SSHL patients, even those who presented with no vestibular symptoms. Patients with pure vertical, pure torsional, gaze evoked nystagmus, or disorders in pursuit or optokinetic nystagmus were also excluded. Torsional nystagmus induced by the Dix-Hallpike maneuver was not observed in any patient. The presence or absence of torsional nystagmus was examined using an infrared charge-coupled device camera.

A total of 127 patients, or 86 patients with low-tone and 41 with high-tone hearing loss, were examined in this study. Low-tone and high-tone SSHL patients were divided on the basis of audiogram patterns according to the criteria indicated in Table 1. We observed no significant difference in age or sex between the 2 audiometric types. In most patients, corticosteroids were administered intravenously as an initial treatment. The recurrence rate of hearing loss and the progression rate to definite Ménière's disease were assessed. We defined recurrence as a second attack of hearing loss with or without vertiginous symptoms. Definite cases of Ménière's disease were diagnosed according to the criteria proposed by the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology–Head and Neck Surgery (15). The follow-up period varied from 6 to 210 months, with a mean of 67 months.

TABLE 1. *Patient selection based on audiogram patterns*

1. Unilateral low-tone type	The average hearing level at 125, 250, and 500 Hz is at least 10 dB worse than that at 2, 4, and 8 kHz
2. Unilateral high-tone type	The average hearing level at 2, 4, and 8 kHz is at least 10 dB worse than that at 125, 250, and 500 kHz
3. No peak and trough	There is a less than 10-dB difference in hearing level at 1 kHz in comparison with that at both the adjacent frequencies—500 Hz and 2 kHz

Low-tone type: 1 and 3.
High-tone type: 2 and 3.

TABLE 2. *Recurrence rate of hearing loss and the rate of progression to definite Ménière's disease*

		Recurrent course			Progression to Ménière's disease		
Initial diagnosis	n	−	+		−	+	
SSHL with vertigo	19	15	4	NS	16	3	NS
SSHL without vertigo	108	72	36		102	6	

NS indicates no significant difference; SSHL, sudden sensorineural hearing loss.

Eye movements were recorded by the ENG technique and sampled at 100 Hz with an A-D converter on a personal computer (16). The presence or absence of SN and the results of caloric testing were reviewed. For detection of SN, subjects were seated in an upright position with the eyes closed for the first 30 seconds. With their eyes still closed, they were instructed to perform simple calculations for 30 seconds. Electronystagmography was performed in most of the patients within 2 weeks of the initial diagnosis. Electronystagmography records were reviewed by 2 of the authors (M.J., H.F.). Nystagmus was considered to be present when 5 or more consecutive nystagmus beats were identified.

Caloric stimuli consisted of alternate irrigation for 60 seconds with 6 L/min of cold and hot air (24 and 50°C). The fast phases of nystagmus were removed to obtain slow-phase eye velocity records. Asymmetry of vestibular function was calculated using the Jongkees formula. Caloric paresis was defined by a response difference of 20% or more between the ears.

Statistical evaluations were performed using the χ^2 and Fisher's exact test. A difference was considered statistically significant at $p < 0.05$. Bonferroni correction was performed to adjust for multiple comparisons.

RESULTS

Of the 127 patients, 40 had recurrent hearing loss. The average time to recurrence for these 40 patients was

TABLE 3. *Recurrence rate of hearing loss and the rate of progression to definite Ménière's disease*

Initial diagnosis	n	Recurrent course		Progression to Ménière's disease	
SSHL with vertigo					
Low tone	4	25.0% (1 of 4)	NS	25.0% (1 of 4)	NS
High tone	15	20.0% (3 of 15)		13.3% (2 of 15)	
SSHL without vertigo					
Low tone	82	40.2% (33 of 82) [a]		6.0% (5 of 82)	NS
High tone	26	11.5% (3 of 26)		3.8% (1 of 26)	

[a]Significant difference after Bonferroni correction.
NS indicates no significant difference; SSHL, sudden sensorineural hearing loss.

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TABLE 4. *Spontaneous nystagmus on ENG*

Initial diagnosis	N	Nystagmus			Detection ratio, %
		Present	Absent		
SSHL with vertigo	19	15	4	[a]	78.9
SSHL without vertigo	108	52	56		48.1

[a]Significant difference at $p < 0.05$.

ENG indicates electronystagmography; SSHL, sudden sensorineural hearing loss.

18.2 ± 26.1 months. Approximately half of them had a recurrence within 6 months from the first episode of hearing loss. There was no significant difference in the recurrence rate of hearing loss when patients were classified with regard to the awareness of vertigo at the onset of hearing loss (Table 2). However, among patients without vertigo, the recurrence rate was significantly higher for those with low-tone than for those with high-tone SSHL (Table 3). The progression rate to definite Ménière's disease was slightly higher in the patients without vertigo than in those with vertigo, but the difference was not statistically significant (Table 2).

We observed SN in patients with vertigo but also in half of the patients without vertigo at the onset of hearing loss (Table 4). Spontaneous nystagmus was easily detected when the patients were asked to perform simple calculations. The direction of nystagmus was independent of the affected side. In the caloric test, 31.6% of patients without vertigo and 22.2% of those with vertigo showed caloric paresis. These data imply that the vestibular system is highly involved even if patients present only cochlear symptoms at the first episode.

Table 5 shows the recurrence rate of hearing loss and the progression rate to definite Ménière's disease when patients were classified with regard to the presence or absence of SN. We observed no significant difference in the recurrence rate of hearing loss between the 2 groups with and without SN. However, among patients with SN, the recurrence rate was significantly higher for those with low-tone SSHL than for those with high-tone SSHL (Table 6). Among the patients with low-tone SSHL without vertigo and without nystagmus, the recurrence rate was 28.6%. In those with nystagmus, the recurrence rate was 52.5%. Some patients with SN subsequently developed definite Ménière's disease, whereas patients without SN at the initial ENG never developed Ménière's (Table 5).

TABLE 5. *Recurrence rate of hearing loss and the rate of progression to definite Ménière's disease*

Initial diagnosis	n	Recurrent course			Progression to Ménière's disease		
		−	+		−	+	
SSHL with SN	67	42	25	NS	58	9	[a]
SSHL without SN	60	45	15		60	0	

[a]Significant difference at $p < 0.05$.

NS indicates no significant difference; SN, spontaneous nystagmus; SSHL, sudden sensorineural hearing loss.

TABLE 6. *Recurrence rate of hearing loss and the rate of progression to definite Ménière's disease*

Initial diagnosis	n	Recurrent course		Progression to Ménière's disease	
SSHL with SN					
Low tone	43	51.2% (22 of 43)	[a]	14.0% (6 of 43)	NS
High tone	24	12.5% (3 of 24)		12.5% (3 of 24)	
SSHL without SN					
Low tone	43	27.9% (12 of 43)	NS	0.0% (0 of 43)	NS
High tone	17	17.6% (3 of 17)		0.0% (0 of 17)	

[a]Significant difference after Bonferroni correction.

NS indicates no significant difference; SN, spontaneous nystagmus; SSHL, sudden sensorineural hearing loss.

DISCUSSION

Recording of SN and caloric tests is widely used to estimate vestibular function. Spontaneous nystagmus reflects static imbalances in the tonic levels of vestibular activity (17). As expected, a high rate of SN was observed in the vertigo-accompanied patients in this study. However, SN was also observed in half of the patients who were not aware of vertigo at the onset of hearing loss, regardless of audiometric patterns. During ENG testing, patients were seated in an upright position. Some performed simple arithmetic tasks to provoke SN. We think that such calculation tasks can help in maintain alertness and preserve the vestibuloocular reflex (18–20).

Spontaneous nystagmus is often present even in healthy subjects (21–23). However, Kumar (24) interprets SN in normal subjects as an oculomotor abnormality for which there is no detectable cause. In patients with cochleovestibular symptoms, SN is considered pathologic (24). The frequency of SN varies according to the recording conditions. Nystagmus has been observed in 1 to 25% of patients during ENG recording with the eyes closed (24–26) and in 10% of patients wearing Frenzel glasses (22). In subjects older than 65 years, SN and positional nystagmus are more frequent (27).

In the present study, 15.8% of the vertigo-accompanied SSHL patients developed Ménière's disease. Some of the SSHL patients without vertigo also developed Ménière's disease (6.0% of the low-tone type and 3.8% of high-tone type). In contrast, on initial ENG, 14.0% of the low-tone–type patients with SN and 12.5% of the high-tone–type patients who also had SN subsequently developed Ménière's disease. When SN was not detected in the initial ENG recordings, no patient developed Ménière's disease, regardless of the audiogram patterns. Thus, objective nystagmus findings at an initial examination are useful for predicting the progression to Ménière's disease.

Ménière's disease is characterized by 2 or more episodes of vertigo concomitant with fluctuating sensorineural hearing loss, tinnitus, and a feeling of fullness or

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pressure in the ear (15). Ménière's disease occurs most commonly between the fourth and sixth decades of life, although children and elderly adults are also affected (28,29). The incidence and prevalence of Ménière's disease vary (30). It has been noted that the prevalence of Ménière's disease in women is slightly higher than that in men (30).

In Japan, the prevalence of Ménière's disease is approximately 17 per 100,000 based on national surveys of hospital attendance in 1977, 1982, and 1990. The proportion has been almost constant among the 3 surveys (31). The clinical course of Ménière's disease differs greatly in individuals. Vestibular and cochlear symptoms may not always occur simultaneously, particularly in the early stages of the disease (32,33). Approximately half of patients with Ménière's disease show symptoms of both vertigo and hearing loss, 36% have hearing loss alone, and 14% have vertigo alone at the onset (32).

Histopathologic studies have demonstrated that endolymphatic hydrops appears first in the cochlea, and then in the saccule, utricle, and semicircular canals (34). Our data suggest that vestibular alterations may already exist in the early stages of Ménière's disease even if the patients have cochlear symptoms alone.

Acute low-tone sensorineural hearing loss is a relatively new disease entity distinct from idiopathic SSHL. It was first introduced in 1982 by Abe (1) and has since been extensively studied primarily in Japan (3–6). The pathophysiology of ALHL may be related to cochlear hydrops or another inner ear disorder closely related to SSHL. Long-term observation of the clinical course has shown that some patients show a recurrence of hearing loss and eventually develop Ménière's disease (4). Electrocochleography and the glycerol test suggest that the pathogenesis of ALHL may be similar to the early stages of Ménière's disease in that both are characterized by endolymphatic hydrops with little or no impairment of hair cells (4–6). In this study, we found that the recurrence rate of hearing loss was especially high in the patients with low-tone SSHL and nystagmus. If it is assumed that the pathophysiology of ALHL involves localized endolymphatic hydrops in the cochlea (4), the absence of SN should be considered as a possible exclusion criterion.

The presence of nystagmus may provide important information for clinicians who are asked daily to diagnose vestibular disorders. Electronystagmography, which provides a clear record of eye movements, is the most common method of vestibular laboratory recording (35). Caloric irrigation, rotatory testing, vestibular evoked myogenic potentials, and galvanic body sway tests are integrated into our routine laboratory evaluations to differentiate among the vestibular disorders. However, they may not always be performed on SSHL patients without vertiginous symptoms due to their expense and inconvenience to the patient. As for the recurrence of hearing loss and progression to Ménière's disease, we conclude that brief ENG examination for nystagmus along with ECochG would provide useful information for treatment planning and prognosis in patients with SSHL of unknown origin, even in those who do not present with vertigo.

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