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Original Article

# Sudden Sensorineural Hearing Loss and Facial Palsy in Patients with Vestibular Schwannoma Based on the Population Data of Korea

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**BACKGROUND:** The prevalence of sudden sensorineural hearing loss and facial palsy in patients with vestibular schwannoma and the association of sudden sensorineural hearing loss or facial palsy with vestibular schwannoma were investigated based on the population data of Korea.

**METHODS:** This retrospective study used the Korean National Health Insurance Service data. Patients with vestibular schwannoma and those with a previous history of sudden sensorineural hearing loss or facial palsy were identified based on diagnostic, medication, magnetic resonance imaging, or audiometric codes from 2005 to 2020. The control group was established with propensity score matching. The risk for vestibular schwannoma in patients with a previous history of sudden sensorineural hearing loss or facial palsy was analyzed.

**RESULTS:** There were 5751 patients in the vestibular schwannoma group and 23 004 in the control group. The rate of patients with a previous history of sudden sensorineural hearing loss in the vestibular schwannoma group (25.8%) was significantly higher than in the control group ($P < .0001$), as was the rate of patients with a previous history of facial palsy in the vestibular schwannoma group (4.7%) ($P < .0001$). Previous history of sudden sensorineural hearing loss was a significant risk factor for vestibular schwannoma (hazard ratio = 7.109, 95% confidence interval = 6.696-7.547). Previous history of facial palsy was also a significant risk factor for vestibular schwannoma (hazard ratio = 3.048, 95% confidence interval = 2.695-3.447).

**CONCLUSION:** The prevalence of sudden sensorineural hearing loss or facial palsy was significantly higher in patients with vestibular schwannoma than in those without vestibular schwannoma. Based on the population data of Korea, sudden sensorineural hearing loss and facial palsy were significant risk factors for vestibular schwannoma.

**KEYWORDS:** Vestibular schwannoma, population-based study, sudden sensorineural hearing loss, facial palsy

## INTRODUCTION

Vestibular schwannoma (VS) is the most common benign tumor of the cerebellopontine angle and originates from the Schwann cells of the vestibular nerves.[1,2] It presents with progressive hearing loss, balance dysfunction, and ultimately severe neurologic sequelae due to compression of the brainstem.[2,3] Approximately 10%-20% of patients with VS were reported to have experienced sudden hearing loss in their lifetime.[1,2]

Sudden sensorineural hearing loss (SSNHL) is characterized as sensorineural hearing loss with 30 dB or more within 72 hours in at least 3 consecutive frequencies.[2,3] More than 90% of patients with SSNHL are idiopathic.[1,2,3] The rate of VS in patients who were treated for SSNHL has been reported to range from 1.9% to 10.2%.[2] Facial palsy is a rare preoperative symptom of VS, occurring in less than 3% of patients with VS, even though large- or medium-sized VS could compress the facial nerve.[4]

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There have been several studies elucidating the association of SSNHL or facial palsy to VS. The rate of SSNHL or facial palsy in patients with VS was higher than the incidence reported in the general population. However, no studies have been conducted regarding the associations compared with a control group based on the population data of the entire country.

Due to the higher possibility of VS in patients with SSNHL or facial palsy, the necessity and benefit of further evaluations such as magnetic resonance imaging (MRI) in those patients should be considered. In this regard, an investigation about the association of SSNHL or facial palsy with VS using population data may be helpful. In this study, the prevalence of SSNHL and facial palsy in patients with VS and the association of SSNHL or facial palsy with VS were investigated based on the population data of Korea.

## MATERIAL AND METHODS

### Subjects

This retrospective study used the Korean National Health Insurance Service data. The results of the present study represent the entire Korean population because all Korean people join the National Health Insurance Service.

Patients who were diagnosed with D33.3 (benign neoplasm of brain and other parts of central nervous system, cranial nerves) or D43.1 (neoplasm of uncertain or unknown behavior of brain and central nervous system, infratentorial brain) in the International Classification of Diseases (ICD) were recruited for the VS group from 2005 through 2020. Among them, patients who had performed brain or temporal MRI and audiometric tests such as auditory brainstem response, pure tone audiometry, or speech audiometry 1 year prior to the first diagnosis with D33.3 or D43.1 were established as the VS group. Patients with benign tumor of the meninges or malignant tumor of the brain and meninges (D32 [benign neoplasm of meninges], C70 [malignant neoplasm of meninges], or C71 [malignant neoplasm of brain] in the ICD) were excluded.

In the Korean National Health Insurance Service, there is the "Special Exception of Assessment for Patient's Excess" system to reduce the patient burden for expensive medical costs of diseases including cancer, rare intractable diseases, and benign neoplasms of the meninges, brain, and central nervous system such as VS. For accuracy, only patients with the "Special Exception of Assessment" code relevant to VS were included in the VS group. The control group with a 4-fold larger number of subjects was established with propensity score matching so that there were no differences in the distributions of sex, age, and household income level between the patient and the control group.

For SSNHL, patients who had visited outpatient clinics twice or more or were admitted once or more with a prescription for steroids under the diagnostic code of H91.2 (sudden idiopathic hearing loss) in the ICD from 2005 through 2020 were defined as those with a previous history of SSNHL in the control group, and those before the onset of VS were defined as those with a previous history of SSNHL in the VS group. For facial palsy, patients who visited outpatient clinics twice or more or were admitted once or more with a prescription for steroids under the diagnostic code of G51.0 (Bell's palsy) in the ICD from 2005 through 2020 were defined as those with a previous history of facial palsy in the control group, and those before the onset of VS were defined as those with a previous history of facial palsy in the VS group.

The Institutional Review Board of The Catholic University of Korea Eunpyeong St. Mary's Hospital approved this study (Approval no: PC22ZISI0051). Written informed consent was waived due to the retrospective nature of the study.

### Data Analysis

Statistical analyses were conducted using SAS Enterprise Guide 7.1 running on SAS 9.4 (SAS Institute, Cary, NC, USA). The proportion of patients with a previous history of SSNHL or facial palsy was analyzed between the VS and control groups with chi-square test. The risk for VS in patients with a previous history of SSNHL or facial palsy was analyzed with univariate and multivariate Cox proportional hazard regression models. Age, sex, and household income level were adjusted in the multivariate Cox proportional hazard regression model.

## RESULTS

There were 5751 patients in the VS group and 23004 in the control group. There were no significant differences in sociodemographic distributions between the 2 groups (Table 1).

The rate of patients with a previous history of SSNHL was 25.8% in the VS group and was significantly higher than in the control group (1.7%) ($P < .0001$). The rate of patients with a previous history of facial palsy was 4.7% in the VS group and was also significantly higher than in the control group (1.0%) ($P < .0001$). In univariate Cox regression analyses, previous history of SSNHL was a significant risk factor for VS (hazard ratio [HR] = 7.285; 95% CI = 6.862-7.733). Previous history of facial palsy was also a significant risk factor for VS (HR = 3.632; 95% CI = 3.213-4.106). In multivariate Cox regression analyses, previous history of SSNHL was a significant risk factor for VS (HR = 7.109; 95% CI = 6.696-7.547). Previous history of facial palsy was also a significant risk factor for VS (HR = 3.048; 95% CI = 2.695-3.447) (Table 2).

## DISCUSSION

The symptoms of VS include hearing loss, dizziness, and tinnitus.[2,5,6] Approximately 20%-30% of patients with VS exhibited SSNHL during disease development.[7] It has been reported that 0.8%-47.5% of SSNHL cases were associated with a diagnosis of VS, whereas 3%-26% of patients with VS had a history of SSNHL.[6,8] It has been reported that

### MAIN POINTS

- The prevalence of sudden sensorineural hearing loss and facial palsy in patients with vestibular schwannoma and the association of sudden sensorineural hearing loss or facial palsy with vestibular schwannoma were investigated based on the population data of Korea.
- The prevalence of sudden sensorineural hearing loss or facial palsy was significantly higher in patients with vestibular schwannoma compared to those without vestibular schwannoma.
- Sudden sensorineural hearing loss and facial palsy were significant risk factors of vestibular schwannoma based on the population data of Korea.

**Table 1.** Sociodemographic Characteristics of Vestibular Schwannoma and Control Groups

| | | Vestibular Schwannoma Group (n = 5751) | | Control Group (n = 23 004) | | *P* |
|---|---|---|---|---|---|---|
| | | n | Percent | n | Percent | |
| Age (year) | <19 | 72 | 1.3 | 245 | 1.1 | .7231 |
| | 20-29 | 226 | 3.9 | 928 | 4.0 | |
| | 30-39 | 613 | 10.7 | 2466 | 10.7 | |
| | 40-49 | 1122 | 19.5 | 4488 | 19.5 | |
| | 50-59 | 1736 | 30.2 | 6944 | 30.2 | |
| | 60-69 | 1326 | 23.1 | 5305 | 23.1 | |
| | 70-79 | 584 | 10.2 | 2396 | 10.4 | |
| | >80 | 72 | 1.3 | 232 | 1.0 | |
| | Average | 53.42 ± 13.55 | | 53.35 ± 13.36 | | .1701 |
| Sex | Male | 2456 | 42.7 | 9832 | 42.7 | .9620 |
| | Female | 3295 | 57.3 | 13172 | 57.3 | |
| Household income level | First quartile (lowest) | 1024 | 17.8 | 4096 | 17.8 | .9997 |
| | Second quartile | 1018 | 17.7 | 4072 | 17.7 | |
| | Third quartile | 1314 | 22.9 | 5269 | 22.9 | |
| | Fourth quartile (highest) | 2395 | 41.6 | 9567 | 41.6 | |

**Table 2.** Risk for Vestibular Schwannoma in Patients with Previous History of Sudden Sensorineural Hearing Loss or Facial Palsy

| Previous History | | Vestibular Schwannoma Group (n = 5751) | | Control Group (n = 23 004) | | *P* | Univariate Cox Proportional Regression Analysis | | | Multivariate Cox Proportional Regression Analysis[a] | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | n | Percent | n | Percent | | HR | 95% CI | *P* | HR | 95% CI | *P* |
| SSNHL | No | 4270 | 74.3 | 22 624 | 98.4 | | 1 | | | 1 | | |
| | Yes | 1481 | 25.8 | 380 | 1.7 | <.0001 | 7.285 | 6.862-7.733 | <.0001* | 7.109 | 6.696-7.547 | <.0001* |
| Facial palsy | No | 5483 | 95.3 | 22 779 | 99.0 | | 1 | | | 1 | | |
| | Yes | 268 | 4.7 | 225 | 1.0 | <.0001 | 3.632 | 3.213-4.106 | <.0001* | 3.048 | 2.695-3.447 | <.0001* |

HR: hazard ratio; SSNHL: sudden sensorineural hearing loss.
[a]Age, sex, and household income level were adjusted.
*$P < .05$.

7.7% of patients with VS experienced 2 or more episodes of SSNHL.[6] The recovery rate of SSNHL in patients with VS decreased with each successive occurrence.[6,9]

The incidence of facial palsy in patients with VS has been reported in the range of approximately 2%-6%.[4,10] It was reported that VS patients with preoperative facial palsy had characteristics of older age and large cystic tumors with significant meatal extension compared to those without preoperative facial palsy.[4] Facial palsy tends to occur in larger compressive tumors with a more medial location.[10]

In the present study, patients with a previous history of SSNHL or facial palsy were significantly more frequent in the VS group compared to the control group. Previous history of SSNHL or facial palsy was a significant risk factor for VS. Although the higher rate of SSNHL or facial palsy in patients with VS has been reported, our study revealed a significantly higher prevalence of SSNHL or facial palsy in patients with VS, and that SSNHL and facial palsy were significant risk factors for VS compared with the control group. MRI is the gold standard for diagnosing VS.[1,2,3,7] Considering these results based on the population data and the importance of early diagnosis of VS using imaging evaluation, further evaluations for VS such as MRI should be considered in patients with SSNHL or facial palsy at the discretion of respective clinicians. Especially, further evaluations for VS should be kept in mind in patients with SSNHL because the risk for VS in patients with a previous history of SSNHL was more than twice as high as that in patients with a previous history of facial palsy. However, contrast-related risks should be considered before performing MRI.

Several mechanisms of SSNHL in patients with VS have been proposed including tumor growth acceleration, tumor volume increase due to interstitial fluid pressure, hemorrhage into tumor, inflammatory immune reaction to the tumor, conduction block of the cochlear nerve, vascular compromise, biochemical changes within the inner ear, and endolymphatic hydrops.[5,6,8,11] Facial palsy in VS may be caused by the rapid growth of the tumor, tumor-induced stretching effects, and intratumoral hemorrhage.[4]

There are several limitations to this study using the population data of Korea. First, patients with other benign tumors of the brain or the

cranial nerves might have been included in the VS group while actual patients with VS may have not been reflected in the patient group. Second, the severity, prognosis, and treatment outcomes of SSNHL or facial palsy could not be compared between the VS and control groups because they were difficult to define in the population data with diagnostic and treatment codes. We aimed to verify the high risk for VS in patients with a previous history of SSNHL or facial palsy. Third, the association of tumor size with SSNHL or facial palsy could not be evaluated because the size could not be verified using the population data.

The possibility of inclusion of patients with facial nerve schwannoma, which was defined with the same diagnostic code as VS, in VS group was the major limitation. Facial nerve tumors including schwannomas, hemangiomas, and neurofibromas are rare, comprising less than 1% of all intratemporal mass lesions.[12] Facial nerve schwannoma is the most common primary tumor of the facial nerve even though the estimated prevalence is extremely low.[12,13] Facial nerve schwannoma presents with symptoms of unilateral peripheral facial palsy and hearing loss.[12] Tumors at the cerebellopontine angle or in the internal auditory canal may also be facial schwannomas, meningiomas, or other tumors. However, facial nerve schwannoma is rarer than VS. More than 80% of tumors in this region are VSs.[1] Therefore, most of the VS group seemed to be constituted of patients with VS rather than other tumors. Even if several patients with facial nerve schwannoma had been included in the VS group, facial nerve schwannoma might also be associated with SSNHL or facial palsy considering the similarity of the tumor to VS regarding location and symptoms.

Despite these limitations, this study has significance in that the prevalence of SSNHL or facial palsy in patients with VS and the risk for VS in patients with SSNHL or facial palsy were evaluated using population-based data in Korea.

In conclusion, the prevalence of SSNHL or facial palsy was significantly higher in patients with VS compared to those without VS. Based on the population data of Korea, SSNHL and facial palsy were significant risk factors of VS.

**Ethics Committee Approval:** This study was approved by the Institutional Review Board of The Catholic University of Korea Eunpyeong St. Mary's Hospital (Approval No: PC22ZISI0051).

**Informed Consent:** The requirement for written informed consent was waived by the Institutional Review Board due to the retrospective nature of the study.

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**Declaration of Interests:** The authors have no conflict of interest to declare.

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