What Is the Real Incidence of Vestibular Schwannoma?

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Objectives: To present the incidence of vestibular schwannoma (VS) in Denmark, compare the incidence with that of previous periods, and discuss the real incidence of VS.

Design, Setting, and Patients: Prospective registration of all diagnosed VS in Denmark, with a population of 5.1 to 5.2 million, during the 6 years from January 1996 through December 2001. Incidence in this period was compared with that of 3 previous periods (July 1976 through June 1983 [first period], July 1983 through June 1990 [second period], and July 1990 through December 1995 [third period]).

Results: In the 1996-2001 period, 542 cases of VS were diagnosed, representing a mean incidence of 17.4 VS/1 million inhabitants per year. Of these, 227 tumors underwent operation, 14 underwent irradiation, and 301 were allocated to observation (wait-and-scan policy). One hundred sixty-six tumors were intrameatal. Size of extrameatal tumors was small in 104; medium in 194; large in 68; and giant (>40 mm) in 10. Compared with incidences of 7.8 VS/1 million inhabitants per year in the first, 9.4 VS/1 million inhabitants in the second, and 12.4 VS/1 million inhabitants in the third periods, the incidence for the 1996-2001 period represents an increase to 17.4 VS/1 million inhabitants per year. The mean incidence for the entire 25.5-year period was 11.5 VS/1 million inhabitants per year.

Conclusions: An estimate of a realistic mean incidence of VS depends on the observation period. Our 25.5-year registration of an entire population showed a mean incidence of 11.5 VS/1 million inhabitants per year. However, the latest period registered represents an incidence of 17.4 VS/1 million inhabitants per year, which, combined with a probable further increase of diagnosed tumors in forthcoming years, suggests a realistic incidence of approximately 13 VS/1 million inhabitants per year.

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As in the 3 previous periods,1-3 all 542 patients with newly diagnosed VS in the 6 years from January 1996 to December 2001 were prospectively registered. Most intrameatal and small tumors did not undergo operation, but were allocated to observation (wait-and-scan management) (Table 1). Most giant and large tumors underwent operation by means of the translabyrinthine approach, and a few by means of the suboccipital or middle fossa approach. Radiotherapy was performed in a few cases. After information about the disease was provided to the patient and the advantages and disadvantages of various treatment modalities were discussed, we followed up and respected the patients’ choice of treatment.

### METHODS

As in the 3 previous periods,1-3 all 542 patients with newly diagnosed VS in the 6 years from January 1996 to December 2001 were prospectively registered. Most intrameatal and small tumors did not undergo operation, but were allocated to observation (wait-and-scan management) (Table 1). Most giant and large tumors underwent operation by means of the translabyrinthine approach, and a few by means of the suboccipital or middle fossa approach. Radiotherapy was performed in a few cases. After information about the disease was provided to the patient and the advantages and disadvantages of various treatment modalities were discussed, we followed up and respected the patients’ choice of treatment.

### RESULTS

Compared with the previous 3 periods,1-3 the total number of diagnosed VS had increased to 542 tumors in the fourth period (Table 2). The mean incidence in the fourth period increased to 17.4 VS/1 million inhabitants per year (Figure 1), and the mean number of diagnosed tumors per year increased to 91.

The size of the diagnosed tumors has gradually decreased through the 4 periods (Table 2). In the first period, the giant and large tumors dominated; in the second period, a significant decrease of the giant tumors and an increase of the small tumors occurred (P < .001, χ² test). In the third period, there was a significant decrease of giant tumors and increase of intrameatal and small tumors (P < .001). In the fourth period, there was a highly significant increase of intrameatal tumors and decrease of giant tumors (P < .001). The 166 intrameatal tumors represent nearly one third of all tumors (Table 2), which is an enormous increase, compared with 0.4% in the first and 0.3% in the second periods.

The percentage of giant tumors decreased to 1.8%, a considerable and significant decrease (P < .001) in relation to the first period with 36.3% giant tumors (Table 2).

Mean size of the extrameatal tumors at diagnosis has gradually decreased for each period, from 28 mm in the first period to 16 mm in the fourth period (Figure 2).

### COMMENT

To illustrate the constantly increasing incidence, the VS can be classified into several categories from the epidemiological point of view. The first category is asymptomatic or silent VS, without any subjective or objective symptoms. We do not know the prevalence or incidence of these VS, or the duration of the asymptomatic period. In addition, we do not know whether all asymptomatic VS will become symptomatic with time. We can imagine, however, that all symptomatic VS have had an initial silent or asymptomatic period and that asymptomatic VS represent a pool of tumors that may become symptomatic. Whether this happens depends on the pa-

### Table 1. Management of VS Diagnosed in Denmark Between January 1996 and December 2001

<table>
<thead>
<tr>
<th>Management</th>
<th>Intrameatal</th>
<th>Extrameatal Extension (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Small (1-10)</td>
<td>Medium (11-25)</td>
</tr>
<tr>
<td>Operation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Translabyrinthine</td>
<td>11</td>
<td>27</td>
</tr>
<tr>
<td>Suboccipital</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Middle fossa</td>
<td>4</td>
<td>7</td>
</tr>
<tr>
<td>Radiotherapy</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Wait and scan</td>
<td>148</td>
<td>69</td>
</tr>
</tbody>
</table>

Total 166 104 194 68 10 542

Abbreviation: VS, vestibular schwannoma.
*Data are expressed as number of tumors.

All demographic data about the patient, symptoms, and tumor size were registered and stored in a database.
asymptomatic VS. Many other factors. Some asymptomatic VS may be discovered incidentally.15

Symptomatic, undiagnosed VS with an unknown prevalence and incidence represent another pool of VS, attributed to the patient’s or the physician’s delay, or both. This pool in Denmark is supposed to have become gradually smaller during the past 25 years, simply because the incidence of diagnosed tumors has increased (Table 2 and Figure 1). This occurred initially because of easier access to computed tomographic scanning and improved quality of the following generations of computed tomographic scanners, and later in the 1980s and 1990s because of the availability of MRI scanning and, later, MRI scanning with gadolinium enhancement. Other important factors are greater awareness on behalf of the patient’s own recognition of the symptoms, which can vary enormously, depending on culture, education, socioeconomic conditions, availability of health care services, and many other factors. Some asymptomatic VS may be discovered incidentally.15

Table 2. Distribution of VS of Various Sizes in Each of the 4 Periods*  

<table>
<thead>
<tr>
<th>Period (No. of VS)</th>
<th>Intrameatal</th>
<th>Extrameatal Extension (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Period (No. of VS)</td>
<td></td>
<td>Small (1-10)</td>
</tr>
<tr>
<td>July 1976 through June 1983 (n = 278)</td>
<td>0.4</td>
<td>13.3</td>
</tr>
<tr>
<td>July 1983 through June 1990 (n = 337)</td>
<td>0.3</td>
<td>21.1</td>
</tr>
<tr>
<td>July 1990 through December 1995 (n = 355)</td>
<td>7.9</td>
<td>29.0</td>
</tr>
<tr>
<td>January 1996 through December 2001 (n = 542)</td>
<td>30.6</td>
<td>19.2</td>
</tr>
<tr>
<td>All 4 periods (n = 1512)</td>
<td>13.0</td>
<td>20.8</td>
</tr>
</tbody>
</table>

Abbreviation: VS, vestibular schwannoma.  
*Data are expressed as percentage of tumors.  
†Because of rounding, percentages may not all total 100.

Figure 1. Mean incidence of vestibular schwannoma (VS) during various periods (newly diagnosed tumors per 1 million inhabitants per year). The parenthetical number below each bar indicates the total number of tumors diagnosed in the corresponding period. Error bars indicate range.

Figure 2. Median size of the extrameatal tumors during the various study periods. Error bars indicate range.

Diagnosed VS has a continuously increasing incidence in each period of up to 17.4 VS/1 million inhabitants per year in the last period (Figure 1). The mean incidence in the entire 25.5 years from July 1976 to December 2001 is 11.5 VS/1 million inhabitants per year, which is an increase compared with the previously published mean incidence of 9.8 VS/1 million inhabitants per year,3 based on the 19.5 years of the first 3 periods from July 1976 to December 1995. The high increase of the incidence in the fourth period to 17.4 VS/1 million inhabitants per year has considerably elevated the mean incidence for the entire 25.5-year period.

We have several indications that the continuous increase of incidence is still due to the diagnosis of previously undiagnosed tumors. Among the first 69 translabyrinthine cases from the first period, the patient’s mean delay in diagnosis was 7 years and the physician’s mean delay was 6 years, for a total mean of 13 years. In the 300 cases that followed, 22% were diagnosed within the first symptomatic year. In the remaining 78% of patients, there was a mean total delay of 7.1 years,16 including the patient’s delay, the physician’s delay, or both. The diagnostic delay became shorter with time, and among 703 patients with VS operated on by translabyrinthine approach, the diagnostic delay was shorter than 1 year in 23%, 1 to 5 years in 43%, and longer than 5 years in 32%. However, no statistically significant differences (x2 test, P>.05) in the length of patient history were found when the patients were grouped consecutively.17 In the most recent period, in which the overall incidence and number of intrameatal tumors has further increased and the extramea-
The increasing high mean incidence reveals the problem concerning tumor growth, ie, what happens with the undiagnosed tumors. Did the patients from the oldest period die with the tumor or because of the tumor? We believe that the former is the case. Because we have been involved in the diagnosis and management of VS for 40 years, we would have noticed if a high number of patients died of a VS at the hospitals, especially as vivisection with opening of the cranium was a routine procedure during the first periods. Therefore, we believe that most of the patients with undiagnosed VS died with the tumor. However, no systematic studies of vivisection records have been performed in our country or reported in the literature.

One intrameatal tumor each was diagnosed during the first and second observation periods, whereas 166 intrameatal tumors were diagnosed in the fourth observation period (Table 2). This indicates that most intrameatal tumors have grown extremely slowly.

**IS THE TRUE INCIDENCE THE MOST RECENT ANNUAL INCIDENCE?**

One may argue that the most accurate incidence derives from the most recent annual incidence, with the most advanced technologies available and easy access to health care, eg, 20 VS/1 million inhabitants per year, as we found in the first year of this survey (2001). This will be the simplest assumption, but then we have to explain what happened to a couple thousand undiagnosed tumors from the previous periods (2530 VS, Table 3). If these tumors are assumed to be growing, they would be diagnosed today as large or giant tumors, and this is not the case (Table 2). If these tumors are not growing, they would all exist today as small tumors without significant symptoms that would lead the patient to a physician, or the patients would have died with or of the tumor. We find this highly unlikely, as argued already.

In our calculation, the highest realistic mean incidence could be 15.3 VS/1 million inhabitants per year. If the assumed incidence is greater than 16%, the number of previously undiagnosed tumors would increase accordingly, stressing the earlier comment even further. Where have these thousands of tumors gone? Have all these patients died with or of the tumor, unacknowledged by our health care system and especially our vivisection pathologists? We believe this is impossible.

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