Increasing annual incidence of vestibular schwannoma and age at diagnosis

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Abstract

During the last 26 years the annual number of diagnosed vestibular schwannomas (VS) has been increasing. The aim of this study is to describe and analyse this increase. Since 1976, 1446 new cases of VS have been diagnosed at the authors' centre. Special focus was on the age at diagnosis, the localization and the size of the tumour. The size of the tumour was registered as either intrameatal or with the largest extrameatal diameter. The annual number of diagnosed VS has increased from 26 in 1976 to 101 in 2001. The size of the diagnosed tumours has decreased from a median of 35 mm in 1979 to 10 mm in 2001. In the first years large and giant tumours dominated, in contrast to recent years in which intrameatal and small tumours dominated. The median age at the time of diagnosis has been almost unchanged through the period (median 55 years). If the decreasing size of the tumour and the increasing incidence of VS can be explained only by earlier diagnosis has been almost unchanged through the median age at the time of diagnosis would decrease simultaneously. In this study, the median age at the time of diagnosis has been almost the 26-year period. This paradox can be explained by the fact that, with easier access to MR scanning, the examination has been offered also to elderly patients, in whom the small and intrameatal tumours dominate.

Key words: Neuroma, Acoustic; Magnetic Resonance Imaging; Age Distribution; Neoplasm Surgery, Incidence

Introduction

In Denmark, with a population of 5.2 million, more than 100 vestibular schwannomas (VS) were diagnosed in 2001, corresponding to an annual incidence of 19.4 VS/million/year.

In the period from January 1957 to July 1976 the incidence of VS in Denmark was estimated to be 5.4 VS/million/year. This estimate was based on publications from Danish neurosurgical departments.¹⁻³ After the introduction of the translabyrinthine approach in Denmark in 1976 a systematic prospective registration of all diagnosed VS has been performed and the mean incidence of VS successively published. In the period from July 1976 to June 1983 the mean incidence was 7.8 VS/million/year.4 In the period from 1983 to 1990 the mean incidence increased to 9.8 VS/million/year,⁵ and in the period from July 1990 to December 1995 to 12.4 VS/million/year.⁶ In the latest period from January 1996 to December 2001 the mean incidence further increased to 17.4 VS/million/year.7

An increase in incidence of VS has also been

reported in Manitoba, Canada, from 5 VS/million/year⁸ during the period 1980–1985, to 13 VS/million/year during the period 1987–1991.⁹ Some other studies¹⁰⁻¹⁴ also found relatively high incidences (10 to 20 VS/million/year) in the 1980s.

The aim of this paper is to describe the increase in number of diagnosed VS through the period, and to relate these findings to the size of the tumours and the age of the patient at the time of diagnosis, in order to assess whether this increase reflects a true increase in annual incidence or whether it is caused by other means (such as easier access to diagnostic tools, computed tomography (CT) and magnetic resonance (MR) scanning examinations, thereby 'fishing a pool' of silent tumours).

Subjects and methods

Since 1976, all patients in Denmark with VS have been referred to one centre, where their data have been entered prospectively into a database. Special note was made of size, localization of the tumour and age of the patient at the time of diagnosis as well as

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TABLE I NUMBER AND SIZE OF VESTIBULAR SCHWANNOMA IN THE DIFFERENT TREATMENT GROUPS

		Size groups						
Treatment groups		Intra- Meatal (n)	1–10 mm (n)	11–20 mm (n)	21–30 mm (n)	31–40 mm (n)	>40 mm (n)	Total
Observed Irradiated Operated	Translab. Middle fossa Sub-occipital	215 3 11 11 -	137 - 121 20 3	133 7 215 - 10	25 4 219 - 5	3 - 126 - 6	- 164 - 8	513* 14** 856 31 32
All operated Total	, I ,	22 240	144 281	225 365	224 253	132 135	172 172	919 1446

*58 patients received surgery later (translabyrinthine:56, middle fossa:1, sub-occipital:1) and six patients received irradiation later **One patient received surgery later (translabyrinthine)

the treatment strategy. The tumours were categorized as either intrameatal or intra- and extrameatal. According to the largest extrameatal diameter at diagnosis the extrameatal tumours were subdivided into small (1–10 mm), medium-sized (11–20 mm), moderately large (21–30 mm), large (31–40 mm) and giant tumours, the latter with an extrameatal diameter larger than 40 mm. This classification follows the recent recommendations on reporting size of VS.¹⁵

During the period from 1976 to December 2001, in total, 1446 patients with unilateral VS have been diagnosed and included. Of these 747 were female and 699 male. The median age at the time of diagnosis was 55 years, range 15 to 84 years.

In contrast to the authors' previous studies, patients with neurofibromatosis 2 (NF 2) are not included in this study due to the different clinical courses of these patients.^{16,17}

The operation was performed primarily in 919 patients, in 856 by the translabyrinthine approach, 31 by the middle fossa approach and 32 by the suboccipital approach (Table I).

Radiotherapy was conducted primarily in 14 patients. One of these patients was operated on later, due to the continuing tumour growth (Table I).

Observation

During the 26 year period 513 patients had been allocated to 'wait and scan' management, with annual MR scanning controls (Table I). The median observation time was three years, range one to 21 years. Forty-six patients in this group had died from non-related reasons during the period. Of the 'wait and scan' group, 58 patients had to receive surgery, and six patients had radiotherapy because of tumour growth (Table I).

The authors' definition of growth for intrameatal tumours was growth to extrameatal extension. In intra- and extrameatal tumours, growth was defined when the tumour had increased more than 2 mm in the largest extrameatal diameter in the period from diagnosis until the last scanning control. In the case of growth the patient was advised to undergo either radiotherapy or surgery.

Results

Annual incidence of diagnosed tumours

Since 1976, the annual number of diagnosed VS has increased almost linearly, from 26 tumours in 1976 to 101 in 2001 (Figure 1), corresponding to an increase in annual incidence from 5.1 VS/million/year in 1976 to 19.3 VS/million/year in 2001. Part of the



Annual number of diagnosed vestibular schwannomas, median age of the patient and median size of the tumour, at the time of diagnosis.



Distribution of different size groups of vestibular schwannomas through the period 1976 to 2001 in 1446 patients.

TABLE II
DISTRIBUTION OF DIFFERENT SIZE GROUPS OF VS THROUGH THE PERIOD 1976 TO 2001

					, ,	Tumor siz	e group						
		Intra	ameatal	1 to	10 mm	11 to	20 mm	21 to	30 mm	31 to	40 mm	>4	0 mm
Year	Total	Ν	%	Ν	%	Ν	%	Ν	%	Ν	%	Ν	%
1976	26	0	0.0	4	15.4	9	34.6	5	19.2	6	23.1	2	7.7
1977	28	0	0.0	9	32.1	8	28.6	2	7.1	0	0.0	9	32.1
1978	37	0	0.0	8	21.6	6	16.2	8	21.6	5	13.5	10	27.0
1979	26	0	0.0	5	19.2	4	15.4	2	7.7	4	15.4	11	42.3
1980	33	1	3.0	4	12.1	9	27.3	8	24.2	5	15.2	6	18.2
1981	30	0	0.0	5	16.7	8	26.7	4	13.3	1	3.3	12	40.0
1982	47	0	0.0	5	10.6	6	12.8	10	21.3	4	8.5	22	46.8
1983	50	5	10.0	7	14.0	10	20.0	15	30.0	3	6.0	10	20.0
1984	32	0	0.0	6	18.8	9	28.1	5	15.6	8	25.0	4	12.5
1985	39	0	0.0	5	12.8	6	15.4	10	25.6	4	10.3	14	35.9
1986	42	1	2.4	7	16.7	11	26.2	10	23.8	4	9.5	9	21.4
1987	43	6	14.0	3	7.0	12	27.9	9	20.9	6	14.0	7	16.3
1988	46	4	8.7	3	6.5	17	37.0	8	17.4	6	13.0	8	17.4
1989	59	6	10.2	12	20.3	12	20.3	12	20.3	8	13.6	9	15.3
1990	50	1	2.0	16	32.0	14	28.0	11	22.0	4	8.0	4	8.0
1991	58	4	6.9	15	25.9	19	32.8	9	15.5	6	10.3	5	8.6
1992	63	10	15.9	12	19.0	15	23.8	11	17.5	7	11.1	8	12.7
1993	48	8	16.7	14	29.2	3	6.3	11	22.9	3	6.3	9	18.8
1994	78	19	24.4	22	28.2	15	19.2	14	17.9	2	2.6	6	7.7
1995	72	16	22.2	13	18.1	27	37.5	13	18.1	3	4.2	0	0.0
1996	82	24	29.3	14	17.1	20	24.4	13	15.9	9	11.0	2	2.4
1997	84	24	28.6	11	13.1	25	29.8	13	15.5	9	10.7	2	2.4
1998	95	28	29.5	18	18.9	18	18.9	19	20.0	10	10.5	2	2.1
1999	83	22	26.5	19	22.9	28	33.7	9	10.8	4	4.8	1	1.2
2000	94	32	34.0	19	20.2	27	28.7	9	9.6	7	7.4	0	0.0
2001	101	29	28.7	25	24.8	27	26.7	13	12.9	7	6.9	0	0.0
Total	1446	240		281		365		253		135		172	

explanation for this increase is the significant increase in the number of intrameatal and small tumours (1–10 mm), from four in 1976 to 54 in 2001 (Figure 2/Table II). The first intrameatal tumour was diagnosed in 1980; this number has increased to 29 in 2001. The percentages of small and medium-sized tumours also increased, while the number of large tumours was almost unchanged throughout the period. In fact the annual number of giant tumours decreased drastically, from 22 in 1982 to one tumour in 1999. Since then, no giant tumours have been detected (Table II).

Median size of diagnosed tumours

Throughout the period, the size of the tumours, at the time of diagnosis, has decreased drastically, from a median size of 35 mm in 1979 to 10 mm in 2001 (Figure 1). At the beginning of the period no intrameatal tumours were diagnosed; in contrast, large and giant tumours constituted 31 per cent of all the tumours. At the end of the period intrameatal tumours dominated (29 per cent), in contrast to the large tumours, which decreased to constitute seven per cent of the tumours in 2001 (Table II).

Incidence of intrameatal VS and number of MR scanners

The increase in the number of MR scanners in Denmark is in accordance with the enormous increase in number of diagnosed intrameatal tumours (Figure 3). The first MR scanner was introduced in 1985 and was the only one in use until 1989. In 2001 there were 29 MR scanners in Denmark, thus, with a population of 5.2 million, resulting in a 'scanner density' of 179.310 inhabitants per MR scanner. From Figure 3 it appears that for each new MR scanner one intrameatal tumour is diagnosed annually.

Age of the patients at diagnosis of VS

Since 1976 the median age of the patients at the time of diagnosis of VS has been almost unchanged, with a slight increase (Figure 1).



Annual number of diagnosed intrameatal vestibular schwannoma and cumulated number of MR scanners.

TABLE III
distribution across different age groups of vestibular schwannoma through the period 1976 to 2001

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					Differen	it age group)				
		<= 40) years	41 to 5	50 years	51 to 6	0 years	61 to 7	0 years	>70	years
Year	Total	N	%	N	%	Ν	%	N	%	N	%
1976	26	3	11.5	6	23.1	12	46.2	4	15.4	1	3.8
1977	28	8	28.6	8	27.6	6	20.7	6	20.7	0	0.0
1978	37	11	29.7	8	21.6	8	21.6	9	24.3	1	2.7
1979	26	6	23.1	8	30.8	3	11.5	6	23.1	3	11.5
1980	33	5	15.2	8	24.2	10	30.3	9	27.3	1	3.0
1981	30	4	13.3	2	6.7	11	36.7	11	36.7	2	6.7
1982	47	12	25.5	7	14.9	14	29.8	11	23.4	3	6.4
1983	50	8	16.0	5	10.0	16	32.0	17	34.0	4	8.0
1984	32	7	21.9	7	21.9	5	15.6	13	40.6	0	0.0
1985	39	9	23.1	7	17.9	9	23.1	11	28.2	3	7.7
1986	42	3	7.1	7	16.7	12	28.6	14	33.3	6	14.3
1987	43	8	18.6	13	30.2	4	9.3	14	32.6	4	9.3
1988	46	10	21.7	5	10.9	10	21.7	13	28.3	8	17.4
1989	59	10	16.9	12	20.3	13	22.0	16	27.1	8	13.6
1990	50	8	15.7	9	17.6	17	34.0	12	23.5	4	7.8
1991	58	8	14.0	16	28.1	10	17.5	14	24.6	10	17.2
1992	63	7	11.1	19	30.2	10	15.9	15	23.8	12	19.0
1993	48	8	16.3	11	22.9	11	22.4	10	20.4	8	16.3
1994	78	11	14.1	13	16.7	24	30.8	19	24.4	11	14.1
1995	72	9	12.5	14	19.4	25	34.7	17	23.6	7	9.7
1996	82	9	11.1	21	25.6	21	25.9	19	23.5	12	14.8
1997	84	15	17.9	21	25.3	22	26.5	18	21.7	8	9.6
1998	95	12	12.5	24	25.0	33	34.4	15	15.6	11	11.6
1999	83	10	12.0	14	16.9	21	25.3	25	30.1	13	15.7
2000	94	12	12.8	14	14.9	34	36.2	22	23.4	12	12.8
2001	101	18	18.0	14	13.9	27	26.7	30	29.7	12	12.0
Total	1446	231		293		388		370		164	

Analysis of the distribution of the patients with VS in various age groups shows that the number of patients aged 40 years or less has been almost unchanged throughout the entire 26-year period (Figure 4/Table III). The increasing number of VS through the late 1980s and 1990s, is primarily constituted by patients older than 50 years. Because of the increased annual incidence in the 1990s, one should expect predominance of younger patients, not the opposite.

At the beginning of the period 81 per cent of the patients were 60 years or younger and only four were older than 70 years. At the end of the period, 59 per cent of the patients were younger than 60 years and 12 per cent were older than 70 years (Table III).



Distribution across different age groups of vestibular schwannoma through the period 1976 to 2001 in 1446 patients.

Age and tumour size at diagnosis

In the age group of 40 years or younger the median size of VS at diagnosis was 25 mm. The size decreased gradually with increasing age, and was 12 mm in the group of patients 70 years or older (Table IV). Even though the median tumour size decreased through the period (Figure 1), the differences in median size of VS between the youngest age group and the older age groups were still present (Figure 5).

Discussion

This study has demonstrated the constantly and regularly increasing annual number of diagnosed VS through a 26-year period in Denmark. The lowest incidence of 5.1 VS/million/year was found in 1976, the highest, 19.3 VS/million/year, in 2001 (Figure 1). Previously, the authors have calculated the mean incidence in four different periods; increasing from 7.8 VS/million/year in the first period (1976 to 1983) to 17.4 VS/million/year in the last period.⁴⁻⁷ The

TABLE IV

MEDIAN TUMOUR SIZE	RELATED	TO AGE A	AT THE	TIME	OF
	DIAGNOSI	S			

Age group	Ν	Median tumour size
40 years or younger	231	25 mm
41 to 50 years	293	18 mm
51 to 60 years	388	15 mm
61 to 70 years	370	15 mm
Older than 70	164	12 mm



Fig. 5

Change in median vestibular schwannoma size (tendencyline) in the different age groups through the period 1976 to 2001.

mean annual incidence for the entire 26-year period was, however, 11.5 VS/million/year.⁷

It is obvious that the relatively low annual incidence in the 1970s and early 1980s is due to the poorer means of diagnosis then, but the real annual incidence is not known. A further increase in the annual incidence may be estimated, but even with an increase to 25 VS/million/year during the following 10-year period, the realistic mean annual incidence for the entire 35 and a half-year period, from July 1976 to December 2011, will not be higher than 15.3 VS/million/year.

In accordance with the increasing annual incidence, this paper demonstrates a constantly decreasing median size at diagnosis of VS (Figure 1). The decrease in median size, from 35 mm in the late 1970s to 10 mm in 2001, is caused by earlier diagnosis. This is influenced by several factors, such as awareness of the patients, general practitioners and otolaryngologists, better audiological testing, and easier access to the first generation of CT scanners. In the 1980s the CT scanners improved further. The result of easier access to better CT scanning and the introduction of MR scanning was the constant decrease in diagnosed giant VS, from 22 in 1982 to 0 in 2000 (Table II). Among the authors' first 59 translabyrinthine cases the patient's own delay in visiting the ENT specialist was seven years, the mean doctors' delay to diagnosis of VS was six years, in total a 13-year delay. In the following series the mean diagnostic delay gradually and constantly decreased.¹⁸ In the authors' 703 translabyrinthine cases the mean diagnostic delay was shorter than one year in 25 per cent of the cases, one to five years in 43 per cent and longer than five years in 32 per cent.¹⁹ Glasscock et al.²⁰ also reported a drastically shorter duration of symptoms in the last 100 cases compared with the first 100 cases.

In the 1990s, the easier access to MR scanners, especially with gadolinium enhancement, had an enormous influence on diagnosis of intrameatal VS, as shown in this study (Figure 3). Since 1980 the percentage of intrameatal VS has increased from 3 per cent to 29 per cent in 2001 (Table II). The

increase in the annual incidence of VS is mainly attributed to the increasing number of diagnosed intrameatal VS.

- In the recent past the incidence of acoustic tumours has appeared to increase
- This paper analyses the size, the age at diagnosis and the localization of 1446 tumours diagnosed in the last 27 years
- The study concludes that, in this period, the number of tumours diagnosed has increased and the size has diminished as intrameatal and small tumours have come to dominate in elderly patients

Therefore the authors still have good reason to believe that the demonstrated continuous increase in the annual incidence of VS is caused by better diagnosis and by finding more and more intrameatal and small VS that are either growing very slowly or have even stopped growing. Only in this way can we explain the diagnosis of so many intrameatal VS in the 1990s (Table II). The increased number of intrameatal tumours parallels the increasing availability of MR scanners (Figure 3). Most of the intrameatal VS have presumably commenced growth in the 1970s, approximately 20 years ago. The presented analysis of age and size supports this view; the median age has not decreased, despite the increased incidence and decreased median size (Figure 1).

If all unilateral VS belonged to the same population of growing tumours with the same growth rate it should be expected that the intrameatal and small tumours would be found in the youngest patients and that the largest tumours would be found in the oldest patients. This was not the case in this study, where the median tumour size was 25 mm in the youngest age group compared with 12 mm in the oldest age group (Table IV). An explanation for this paradox could be that many of the intrameatal VS grow very slowly within the meatus and remain intrameatal for many years, being found in the oldest age groups. It seems that one population of tumours is growing and another, i.e. most of the intrameatal tumours found in the older age group, does not seem to grow. Many of these tumours may be asymptomatic and found by chance, in an age group in which where hearing loss and dizziness become common, because of the easier access to MR scanning.

Conclusion

With easier access to MR scanning, growing vestibular schwannomas are found earlier and smaller. The increase in annual incidence of diagnosed tumours is mainly explained by the diagnosis of small and intrameatal tumours in the older age group.

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