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Hearing in Patients with Intracanalicular Vestibular Schwannomas

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Key Words
Vestibular schwannoma · Acoustic neuroma · Hearing loss

Abstract
This paper reports data on the spontaneous course of hearing in 156 patients with purely intracanalicular vestibular schwannomas. The mean pure tone average (PTA) was 51 dB HL and the mean speech discrimination score (SDS) 60% at diagnosis. The risk of a significant subsequent hearing loss (≥10 dB PTA or ≥10% SDS) was 54% during 4.6 years of observation. Patients with normal speech discrimination at diagnosis had a significantly smaller risk of loosing hearing. The hearing loss at diagnosis and during observation was not related to age, gender, diagnostic tumor size, tumor-induced expansion of the internal auditory canal or tumor sublocalization (fundus, central or porus). However, the loss of PTA was smaller in shrinking tumors and the PTA deterioration rate correlated with the volumetric tumor growth rate. After 4.6 years observation, the PTA had increased by 14–65 dB HL, and the SDS reduced by 17–43%. The proportion of patients eligible for hearing preservation treatment as determined by word recognition score class I (70–100% SDS) was reduced to 28% (a 44% reduction), and by AAO-HNS class A to 9% (a 53% reduction).

Introduction

An asymmetrical hearing loss is the most frequent symptom leading to the diagnosis of vestibular schwannoma [Harner et al., 2000]. A major part of the patients present with a hearing loss, which is not serviceable with a hearing aid and which excludes a meaningful attempt of hearing preservation by surgery or radiotherapy, typically due to a poor capability of speech discrimination. Some patients do, however, have good hearing at diagnosis and may be candidates for treatment attempting hearing preservation. However, proper assessment of the rate and degree of hearing preservation after surgical or radiotherapeutical intervention is meaningless without knowledge on the spontaneous course of hearing following the diagnosis of these tumors.

The incidence of diagnosed vestibular schwannoma is increasing, due at least in part to an increased number of magnetic resonance imaging (MRI) scanners [Stangerup et al., 2004; Evans et al., 2005]. As MRI allows visualization of smaller tumors than the previously used computer tomography, the increased incidence is primarily a question of an increased detection of small and often exclusively intracanalicular tumors. These tumors do not cause brain stem or adjacent cranial nerve compression, and unless significant growth occurs, the only reasonable cause for treatment is attempted hearing preservation.

The spontaneous course of hearing following the diagnosis of a purely intracanalicular vestibular schwanno-

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ma is largely unknown and may in theory depend on several factors, e.g. sublocalization within the canal, growth of the tumor or tumor-induced expansion and increase in the pressure in the internal auditory canal (IAC) [Lapsiwala et al., 2002]. The first aspect may further be decisive for the choice of surgical approach, as a tumor in the fundus of the canal is readily accessible through the middle fossa approach, whereas a central or porus-near tumor can be reached via both the middle fossa and retrosigmoid approach.

This paper reports data on the spontaneous course of hearing in 156 patients with purely intracanalicular vestibular schwannomas primarily allocated to observation by repetitive MRI. The hearing at and following diagnosis is related to tumor size, sublocalization, volumetric growth pattern and expansion of the IAC. The patient group represents all patients diagnosed and prospectively registered with a sporadic, unilateral and exclusively intracanalicular vestibular schwannoma in Denmark during the period 1976–2004, and with at least a diagnostic and one follow-up audiogram and MRI available.

Subjects and Methods

All patients diagnosed with a tumor in the IAC or/and the cerebellopontine angle resembling a vestibular schwannoma have been registered prospectively in a database at one center in Denmark (5.2 million inhabitants) since 1975.

In November 2004, data on all sporadic, unilateral and purely intracanalicular tumors were drawn from the base, yielding 325 patients. Fifty-two patients diagnosed by a CT scan were excluded due to the inaccuracy of this type of imaging, leaving 273 patients diagnosed by MRI. All diagnostic and control images on these patients were retrieved and the sublocalization and three-dimensional size of the tumor determined by measurement. In addition, the images were analyzed for tumor-induced expansion of the IAC (tumor filling the radial aspect of an enlarged canal, compared to the contralateral side). A control MRI was not available in 61 patients (22 patients died before the first follow-up MRI, a control MRI had not been performed in 20 patients and the images from 19 patients could not be retrieved) and one of the three tumor dimensions was not present on the MRI in 16 patients (the coronal projection was missing), yielding 196 patients with at least two sufficient MRI scans for volumetric determination of tumor size, growth frequency and rate.

Most tumors had an ellipsoid shape and the volume was calculated as a product of the three dimensions, using the formula for an ellipsoid: volume = $1/6 \times 3.14 \times d1 \times d2 \times d3$. An increase of at least 2 mm in any diameter was defined as growth and a decrease of at least 2 mm as shrinkage. The absolute and relative volumetric growth rates were calculated.

Tumor sublocalization categories were defined as either fundus near (no CSF between the tumor and the fundus and CSF between the tumor and the porus on T_2-weighted images), central (CSF – or no CSF in tumors filling the entire canal – between the tumor and the fundus and between the tumor and the porus), or porus near (no CSF between the tumor and the porus and CSF between the tumor and the fundus).

The detailed data on tumor size, sublocalization, growth pattern and IAC expansion in these 196 patients have been published recently [Caye-Thomasen et al., 2006].

A diagnostic audiogram was available in 182 of the 196 patients, whereas diagnostic and follow-up audiograms were available in 156 patients. The pure tone average (PTA) was calculated as the mean of the pure tone hearing thresholds at 500, 1000, 2000 and 4000 Hz. Speech audiometry was performed in quiet using standardized word lists read from a CD and the speech discrimination score (SDS) determined by the number of words correctly repeated (in percent) at the most comfortable sound pressure level, according to the masking rules.

Median age of the 156 patients was 57 years at diagnosis (range 15–77) and the male to female ratio was 1.58. The mean observation period between first and last MRI/audiometry was 4.6 years (standard error of mean, SEM, 0.25). The cumulated actual observation was 73% of the ideal observation (fig. 1). Detailed data on size, sublocalization and growth pattern are given in table 1.

Table 1. Various characteristics of the 156 intracanalicular vestibular schwannomas at diagnosis and at the last evaluation

	Diagnostic MRI		Last MRI	
	n	%	n	%
Largest intrameatal diameter, mm				
1–5	31	20	26	17
6–10	101	65	84	54
>10	24	15	18	12
Largest extrameatal diameter, mm				
1–5	–	–	16	10
6–10	–	–	8	5
>10	–	–	4	3
IAC expansion				
Yes	31	20	54	35
No	125	80	102	65
Tumor sublocalization at diagnosis				
Fundus	29	19	–	–
Central	80	51	–	–
Porus	47	30	–	–
Extrameatal extension				
Yes	0	0	28	18
No	156	100	128	82
Volume				
Decreased	–	–	18	12
Unchanged	–	–	71	45
Increased	–	–	67	43
Mean volume	112 mm^3		227 mm^3	

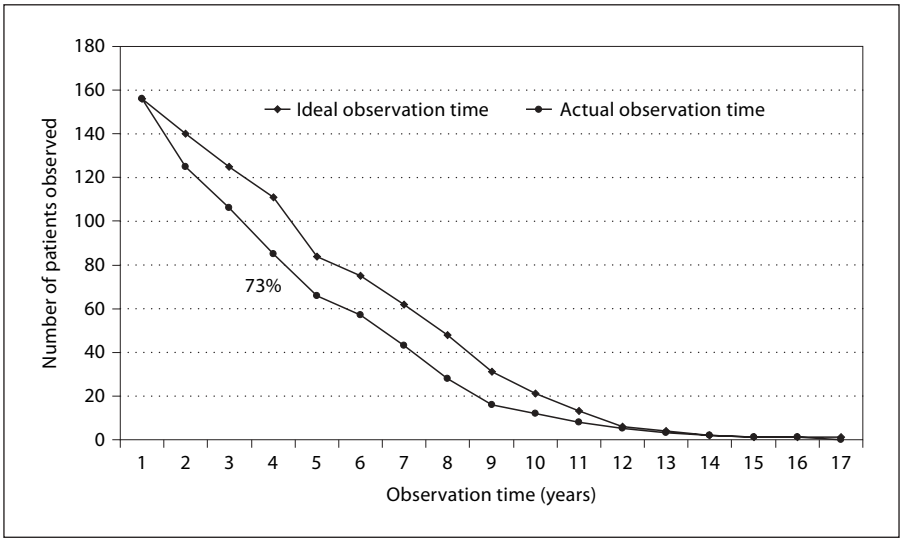

Fig. 1. Actual and ideal observation in 156 patients with intracanalicular vestibular schwannomas.

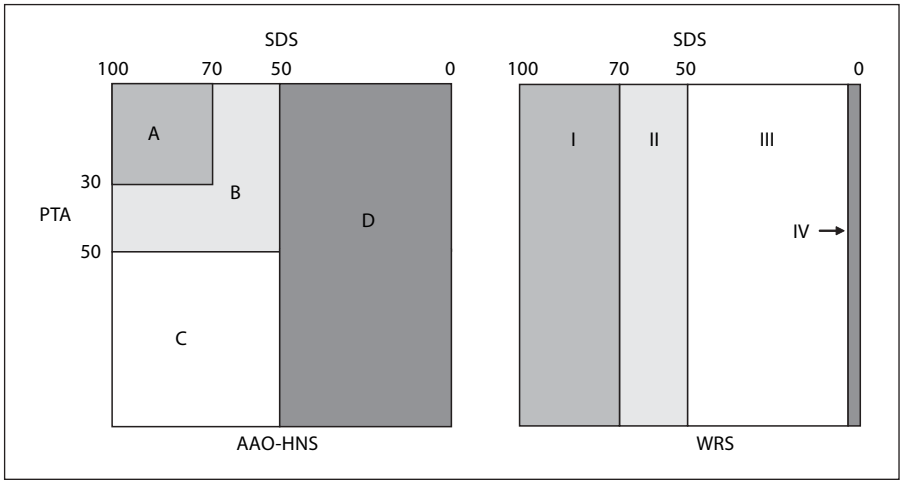

Fig. 2. AAO-HNS and WRS classifications of hearing acuity.

Tumor size, sublocalization, IAC expansion, growth pattern, as well as gender and age were related to the data on PTA and SDS, including classification according to monosyllabic word recognition score (WRS) [Meyer et al., 2006] and American Academy of Otolaryngology – Head and Neck Surgery (AAO-HNS) [1995] (fig. 2).

The nonparametric χ^2, Fisher's exact, Mann-Whitney and Kruskal-Wallis tests were used for comparison of groups. The Spearman rank test (two tailed) and linear regression were used for correlation analyses, and $p < 0.05$ was chosen as the level of significance. Because of the variability of the observation period (high number of censored data), the Nelson-Aalen survival plot was used for the graphic illustration of the cumulated risk of loosing preservable hearing (fig. 3). By using Nelson-Aalen survival statistics, all patients add to the calculation of risk over time, but only with their actual observation period.

Results

Hearing in Intracanalicular Vestibular Schwannomas

The average pure tone audiogram presented with a low-, mid- and primarily high-frequency hearing loss compared to the contralateral ear (fig. 4). The mean PTA was 51 dB HL (SEM 1.8) and the mean SDS was 60% (SEM 2.7) at diagnosis in the tumor ear, compared to significantly lower 20 dB HL (SEM 1.4) and 4% (SEM 0.8) in the contralateral ear ($p < 0.0001$ for both numbers, Mann-Whitney). Overall, a significant increase in PTA and decrease in SDS occurred during observation on the tumor ear ($p < 0.0001$, Mann-Whitney), and PTA increased on the contralateral ear ($p < 0.037$, Mann-Whitney). Fifty percent of the patients had class I WRS and

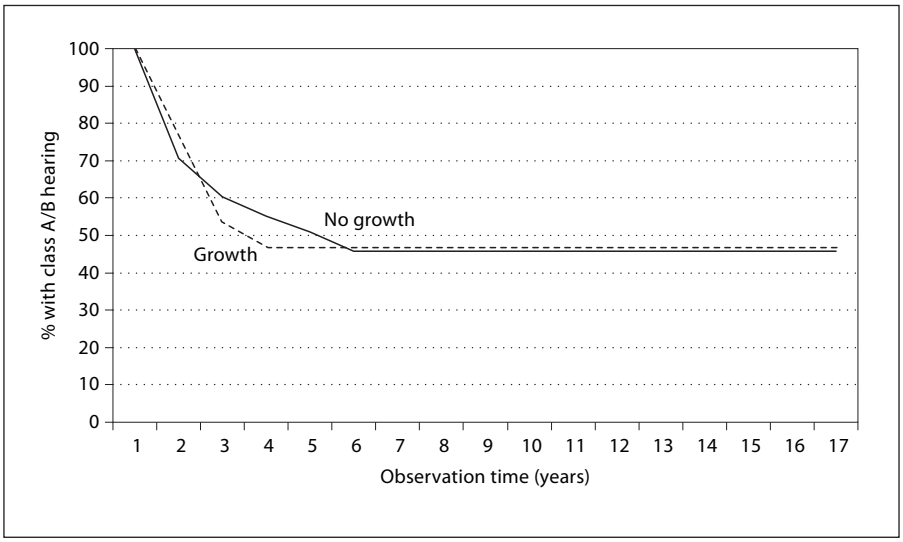

Fig. 3. Percent of patients with AAO-HNS class A/B hearing during observation in growing and nongrowing intracanalicular vestibular schwannomas. Seventy patients (45%) had class A/B hearing at diagnosis.

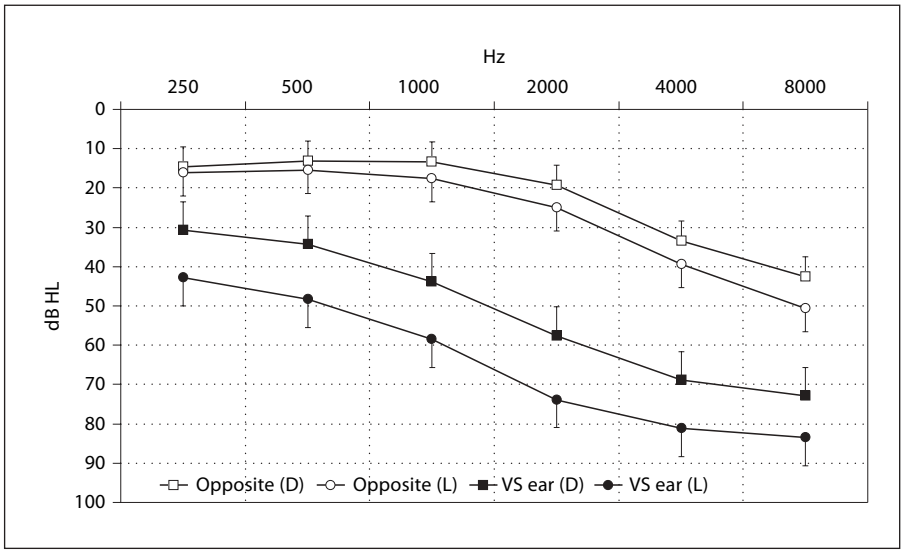

Fig. 4. Average pure tone hearing at various frequencies in the vestibular schwannoma (VS) ear and the opposite (normal) ear at diagnosis (D) and last evaluation (L) (n = 156). The error bars represent the standard error of the mean.

19% had AAO-HNS class A hearing in the tumor ear at diagnosis. During the observation period, these numbers were reduced to 28% (44% reduction) and 9% (53% reduction), respectively (tables 2, 3). The rate and cumulated risk of loosing AAO-HNS hearing class A or B following diagnosis are displayed in figure 3, showing that the hearing is lost during the very first years after diagnosis and that 53% loose class A/B hearing during observation. Compared to the contralateral ear, the cumulated risk for hearing loss during observation was higher for the tumor ear ($p < 0.001$, χ^2). The rate and degree of hearing loss were also higher for the tumor ear ($p < 0.0001$ for both PTA and SDS, Mann-Whitney).

The risk of hearing loss was not related to the diagnostic PTA, which in addition did not correlate with the subsequent hearing deterioration rate.

Patients with Normal Discrimination at Diagnosis

Twenty-six patients (17%) had a normal (100%) SDS in the tumor ear at diagnosis. All of these patients had an SDS above 70% (WRS class I) at the last follow-up (mean 4 years; SEM 1.7). The last mean SDS was 95% (range 80–100, SEM 1.2). The cumulated risk of loosing 10 dB PTA or more was 23% in this group and the cumulated risk of loosing 10% SDS or more was 38%. The equivalent risk for patients with an SDS below 100% at

Table 2. AAO-HNS hearing class distribution in relation to tumor sublocalization at diagnosis and at the last audiogram, in percent

	A	B	C	D
Porus				
At diagnosis	18	24	20	38
At last audiogram	15	12	15	58
Central				
At diagnosis	21	27	14	38
At last audiogram	8	11	16	65
Fundus				
At diagnosis	14	29	34	23
At last audiogram	3	12	55	30
All				
At diagnosis	19	26	20	35
At last audiogram	9	12	23	56

Table 3. WRS class distribution in relation to tumor sublocalization at diagnosis and at the last audiogram, in percent

	I	II	III	IV
Porus				
At diagnosis	44	13	25	13
At last audiogram	29	11	31	29
Central				
At diagnosis	44	18	27	11
At last audiogram	25	10	36	28
Fundus				
At diagnosis	68	9	17	6
At last audiogram	37	30	10	23
All				
At diagnosis	50	15	25	10
At last audiogram	28	14	30	28

Table 4. Size and growth data on the group of growing tumors in relation to tumor sublocalization within the IAC

	Tumors	Mean volume, mm^3		Mean absolute growth mm^3/year	Mean relative growth %/year	IAC expansion, %		Growth to extrameatal extension, %
		first MRI	last MRI			first MRI	last MRI	
Fundus	10	69 (5.1)	219 (11.9)	74 (6.5)	134 (8.9)	20	40	20
Central	38	124 (3.4)	482 (13.6)	112 (7.2)	106 (5.5)	24	61	50
Porus	19	111 (5.0)	300 (12.9)	82 (6.4)	110 (6.0)	5	42	32
All	67	112 (3.1)	391 (9.9)	98 (4.8)	111 (5.1)	18	52	40

Numbers in parentheses are SEM.

diagnosis (n = 130) was a significantly higher 57% for PTA ($p = 0.0033$, χ^2) and had a tendency to be higher for SDS at 59% ($p = 0.096$, χ^2). Fifty-one patients (33%) had an SDS between 70 and 99%. The group with normal SDS did not differ significantly from the rest of the patients with respect to tumor size, sublocalization, IAC expansion, growth pattern or rate.

Tumor Sublocalization, Size and Volumetric Growth Pattern

The sublocalization, size and growth data for the 156 patients with diagnostic and control audiograms are presented in tables 1 and 4, and did not differ significantly in any parameter from the equivalent data found in the original 196 patients outlined in the Materials and Methods section. There were no significant differences in observation time between the different groups of sublocalization and growth pattern. The overall mean absolute growth rate was 41 mm^3/year (SEM 8.1) and the mean relative growth rate 46%/year (SEM 9.2). For the group of shrinking tumors, the mean shrinking rate was 11 mm^3/year (SEM 4.5) and the relative shrinking rate 15%/year (SEM 4.3). The data for growing tumors are presented in table 4.

Hearing in Relation to Tumor SubLocalization and Size at Diagnosis

No significant differences in growth pattern, PTA, SDS, AAO-HNS or WRS hearing class occurred in relation to tumor sublocalization (tables 2–5), and the cumulated risk of hearing loss was not related to either diagnostic sublocalization or size (table 6).

Table 5. Mean PTA and SDS at diagnosis and at the last evaluation, in relation to tumor sublocalization

		Diagnostic audiogram	Last audiogram	p value (Mann-Whitney)
Porus tumors (n = 47)	PTA	51 (3.5)	63 (3.1)	0.04
	SDS	58 (5.2)	45 (5.0)	0.08
Central tumors (n = 80)	PTA	51 (3.0)	60 (2.8)	<0.0001
	SDS	58 (4.9)	40 (5.5)	0.0008
Fundus tumors (n = 29)	PTA	52 (3.2)	65 (3.5)	0.005
	SDS	68 (5.1)	51 (5.2)	0.016
All tumors (n = 156)	PTA	51 (1.8)	65 (2.0)	<0.0001
	SDS	60 (2.7)	43 (3.1)	<0.0001
Contralat. ears (n = 156)	PTA	20 (1.4)	24 (1.2)	0.037
	SDS	96 (0.8)	94 (1.1)	0.14

Numbers in parentheses are SEM.

Table 6. Cumulated risk (%) of loosing ≥10 dB PTA or ≥10% speech discrimination during observation

	All VS ears	Fundus tumors	Central tumors	Porus tumors	Shrinking tumors	Stable tumors	Growing tumors	+IAC	–IAC	All contralat. ears
PTA loss >10 dB	54[a]	47	54	57	28[b]	55[b]	60[b]	45	56	16[a]
SDS loss >10%	52[a]	57	52	49	50	54	51	32	57	9[a]

[a] Significant differences between the tumor ear and the contralateral ear ($p < 0.001$, χ^2, for both PTA and SDS).
[b] Tendency to a smaller risk of loosing PTA for shrinking tumors, compared to stable and growing tumors ($p = 0.053$, χ^2).

Table 7. Mean PTA and SDS at diagnosis and at the last evaluation in relation to tumor growth pattern

		Diagnostic audiogram	Last audiogram	p value (Mann-Whitney)
Shrinking tumors (n = 18)	PTA	60 (3.4)	68 (4.5)	0.14
	SDS	63 (4.4)	50 (4.8)	0.3
Stable tumors (n = 71)	PTA	50 (3.0)	63 (2.8)	0.001
	SDS	61 (4.0)	46 (4.3)	0.007
Growing tumors (n = 67)	PTA	51 (2.9)	67 (3.1)	0.0002
	SDS	58 (3.8)	40 (5.1)	0.002
All tumors (n = 156)	PTA	51 (1.8)	65 (2.0)	<0.0001
	SDS	60 (2.7)	43 (3.1)	<0.0001
Contralat. ears (n = 156)	PTA	20 (1.4)	24 (1.2)	0.037
	SDS	96 (0.8)	94 (1.1)	0.14

Numbers in parentheses are SEM.

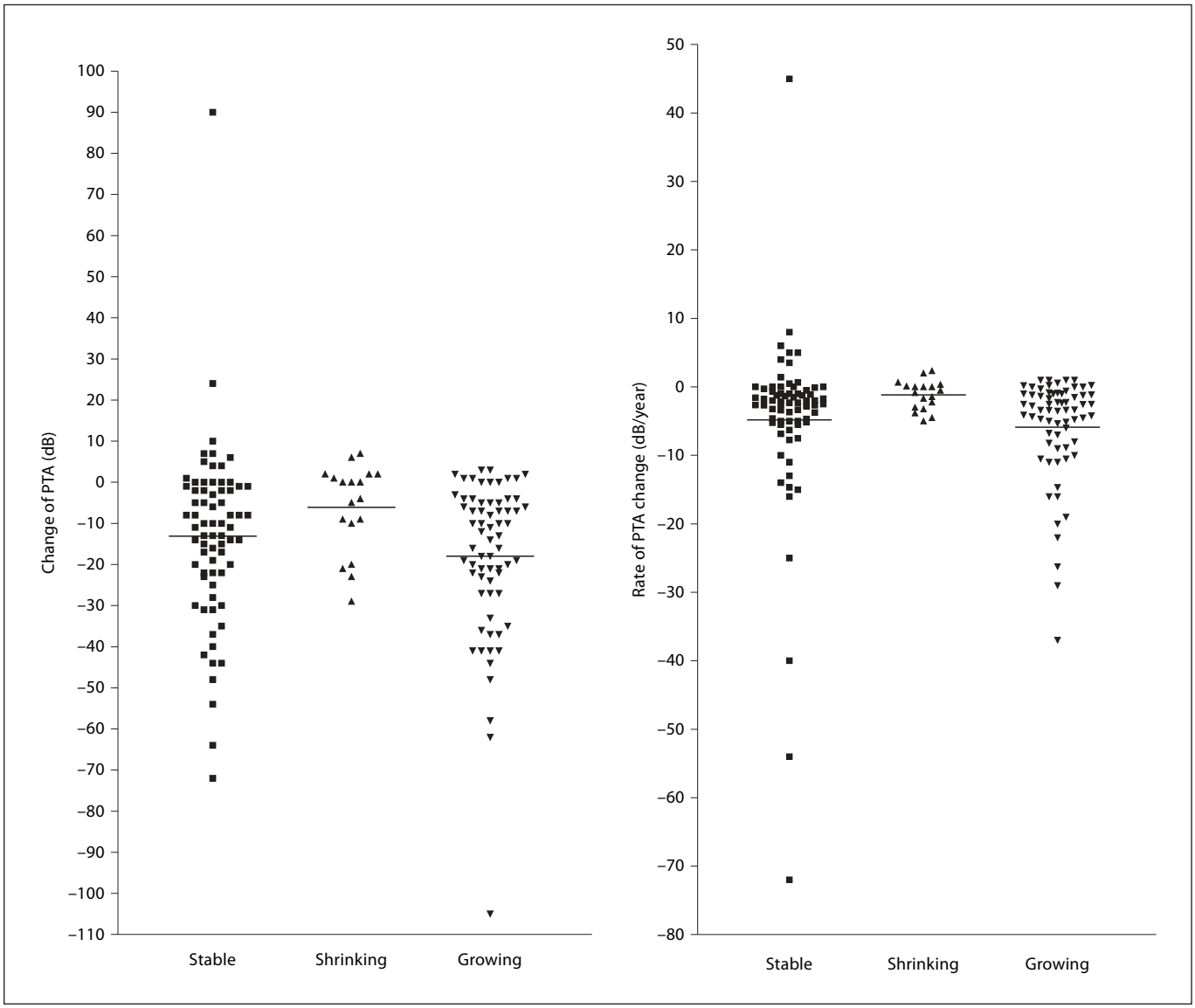

Fig. 5. Scattergram and mean (horizontal bar) of the rate and total change of the PTA during observation of 156 intracanalicular vestibular schwannomas grouped in relation to growth pattern. p = 0.028 for change of PTA and p = 0.01 for rate of PTA change (both Kruskal-Wallis test). The rate and degree of PTA loss is significantly lower for the group of shrinking tumors.

A weak correlation existed between both the diagnostic and last tumor size, and the average PTA deterioration rate (loss of PTA in dB/year; r = –0.17 and –0.26, respectively). However, linear regression analyses showed no significant deviation from zero in both instances (p = 0.72 and 0.27, respectively). No other correlations existed.

Hearing in Relation to Tumor Growth Pattern

The observational hearing deterioration was significant overall for the group of stable tumors and growing tumors, but not for the group of shrinking tumors (table 7). In addition, the average pure tone hearing deterioration rate and the mean loss of PTA during observation were greater for the stable and growing tumors, compared to the shrinking tumors (fig. 5). The difference be-

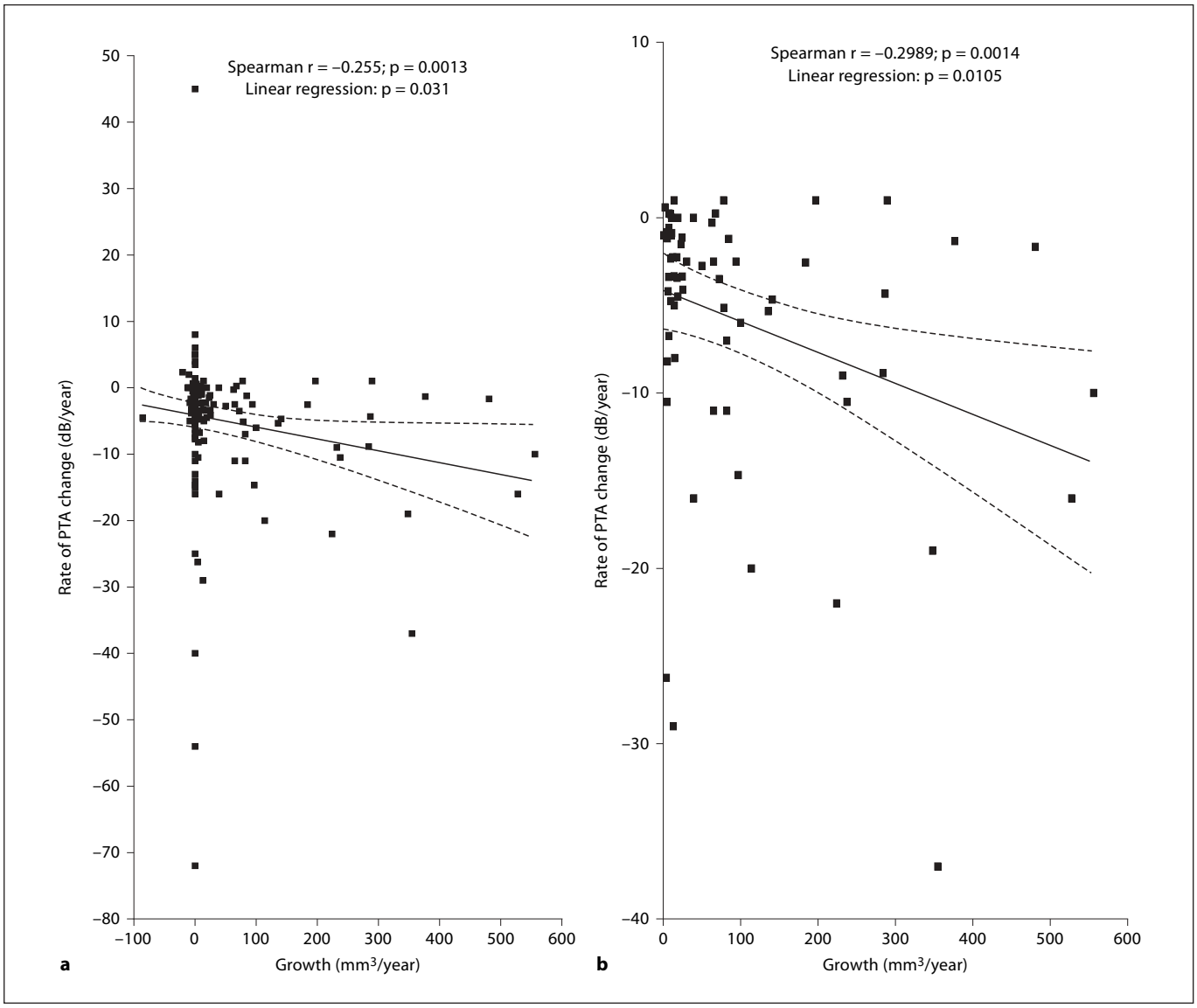

Fig. 6. Scattergram showing the correlation between the PTA deterioration rate and the growth rate in the total patient material (**a**) (n = 156) and in the group of growing tumors (**b**) (n = 67).

tween the group of stable and growing tumors was not significant. However, grouping the shrinking with the stable tumors, the growing tumors did have a significantly higher deterioration rate [5.88 dB/year (SEM 0.9) vs. 4.1 dB/year (SEM 0.7); p = 0.043, Mann-Whitney] and a tendency to a greater mean loss of the PTA [18 dB (SEM 2.3) vs. 12 dB (SEM 1.9); p = 0.068, Mann-Whitney]. Similarly, the average rate and mean loss of PTA was greater for the group of growing tumors extending into the cerebellopontine angle, compared to all tumors remaining within the IAC during observation (p = 0.0001 and p = 0.006, respectively, Mann-Whitney). Correlation analyses showed that the PTA deterioration rate did indeed correlate positively with the absolute growth rate (fig. 6), but not with the relative growth rate. No other correlations existed. Specifically, the loss of speech discrimination was not related to any characteristic of growth.

The cumulated risk of hearing loss was not related to growth occurrence, although there was a tendency to a lower risk for shrinking tumors (p = 0.053; χ^2, table 6).

No relation existed between any hearing parameter and IAC expansion.

Hearing in Relation to Age and Gender

No relation existed between any hearing parameter and gender or age.

Discussion and Conclusion

From this study on the hearing acuity in 156 patients with purely intracanalicular vestibular schwannomas, we can conclude that the mean PTA on the tumor ear is 51 dB HL and the mean SDS is 60% at diagnosis, compared to 20 dB HL and 96% on the contralateral ear. Fifty percent have class I WRS and 19% have AAO-HNS class A hearing. During the following observation of the tumor, the hearing loss progresses at a faster rate than in the contralateral, healthy ear. The progression occurs primarily within the very first years after diagnosis. At a mean of 4.6 years after diagnosis, the PTA is increased by 14 dB to 65 dB HL, and the SDS reduced by 17–43%. The proportion of patients with WRS class I is reduced to 28% (44% reduction) and with AAO-HNS class A to 9% (53% reduction). Patients with normal speech discrimination at diagnosis had a significantly smaller risk of a subsequent hearing loss (see below). The risk, rate and degree of hearing loss in the tumor ear is not related significantly to age, gender, IAC expansion, tumor sublocalization or size. However, the rate and degree of pure tone hearing loss is correlated positively to the absolute volumetric tumor growth rate. As an extension of this finding, the 18% of the tumors that eventually expand into the cerebellopontine angle have a faster rate and degree of pure tone hearing loss compared to the tumors remaining within the canal. In addition, the loss of PTA is significantly smaller in the 12% of tumors displaying shrinkage during observation, compared to the stable and growing tumors. As specified, these correlations only exist for the pure tone hearing, whereas no characteristics of growth relates to the ability to discriminate speech.

The Cause of Hearing Loss

As the absolute volumetric growth rate correlates with the hearing deterioration rate, it seems a fair assumption that tumor pressure on the cochlear nerve induces the hearing loss. This assumption is supported by a study of 20 patients with both an intra- and extrameatal tumor component, as the wave V latency in measuring auditory evoked potentials correlated with the pressure in the IAC, as measured during surgery through the retrosigmoid approach [Lapsiwala et al., 2002]. However, Badie et al. [2001] found that the IAC pressure correlated with the amount of tumor in the IAC in 15 patients, but could not document a significant relation between the pressure and hearing function , and Nadol et al. [1996], observed no correlation between lateral extent of IAC invasion and PTA/SRT scores in 75 patients. In addition, Odabassi et al. [2002] showed that the extent of IAC involvement was not related to the negative effects of the tumor on cochlear function as represented by DPOAEs. In the present material, no relation existed between any parameter of hearing and expansion of the IAC. Thus, the overall findings of these studies suggest that cochlear nerve compression is definitely not the only cause of hearing loss in vestibular schwannoma, leaving other causes, e.g. vascular compromise, abnormal inner ear fluid composition and cochlear hair cell loss, as subjects of speculation [Prasher et al., 1995; Kobayashi et al., 1996].

Bias of Hearing and Growth Data

The main strength of the present data is the prospective and consecutive one-center registration of all patients diagnosed with vestibular schwannoma during the period 1976–2004 in Denmark, with a population of 5.2 million inhabitants. The data are thus without patient referral bias. As all the present patients diagnosed with a purely intracanalicular vestibular schwannoma by MRI have been allocated primarily to observation, the material is in addition without patient selection bias. The referral and patient selection bias is a problem in a vast majority of previously published papers, and this may in conjunction with small sample sizes be the main explanation for the variability of reported growth rates and hearing levels.

Our data are, however, biased by the fact that patients diagnosed by a CT scan were excluded due to the inaccuracy of this type of imaging. Furthermore, a control MRI was not available in 61 patients (22 patients died before the first follow-up MRI, a control MRI had not been performed in 20 patients and the images from 19 patients could not be retrieved). Lastly, one of the three tumor dimensions was not accessible in 16 patients, as the coronal MRI projection was missing. Of the remaining 196 patients with at least two MRIs, a diagnostic and at least one follow-up audiogram was retrievable in 156 (80%). An ideal follow-up of these patients would be to perform an MRI and an audiogram on the day of retrieving data from the database. Compared to this hypothetical situation, our follow-up was 73% (fig. 1).

Comparison with Previously Published Data

All studies on the hearing and growth pattern of vestibular schwannoma, except the study by Stangerup et al. [2006a], have been on highly selected patients, as these typically were observed only because they were unfit to go through surgery due to old age or concurrent disease [Nadol et al., 1996; Yamamoto et al., 1998; Warrick et al., 1999; Haapaniemi et al., 2000; Walsh et al., 2000; Massick et al., 2000; Tschudi et al., 2000; Rosenberg, 2000; Sakamoto et al., 2001; Graamans et al., 2003; Grayeli et al., 2005].

The largest previous sample of observed purely intracanalicular tumors is that of Graamans et al. [2003], who followed 19 patients with nongrowing tumors for 7 years. A progressing hearing loss was without correlation with tumor size, which is in agreement with our findings. Massick et al. [2000] followed 13 patients prospectively for an average of 3.8 years, and found a correlation between volumetric growth and deterioration of both the PTA and SDS (8 growing tumors). While the relation between growth and deterioration of PTA was confirmed in our material, we could not relate any parameter of growth to change of SDS.

Sakamoto et al. [2001] followed 31 mixed tumors (intra- and extrameatal) for 33 months and found a correlation between a mean annual, one-dimensional growth rate of 2.4 mm and an average PTA loss of 2.3 dB/year, although no regression analysis was performed. No correlation was found between growth rate or hearing loss rate and age, size or PTA at diagnosis. Grayeli et al. [2005] reported similar findings in a mixed material, which are equivalent to our results, strengthened by the volumetric determination of size and growth.

As in the present patient sample, Walsh et al. [2000] found no audiological factors predictive of growth in 8 intracanalicular tumors observed for 44 months. However, the risk of hearing loss was higher for the growing tumors, as the risk of loosing AAO-HNS class A/B hearing was 80% for the growing and 14% for the stable tumors. This is, however, a very small sample of patients and this huge difference in risk related to growth pattern is not in agreement with our observations (table 6). Rosenberg [2000] reported the risk of loosing >10 dB PTA to be 51% and the risk of loosing >15% SDS to be 38% in 45 mixed tumors, followed for around 4 years (not specified). Stangerup et al. [2006a], who employed two-dimensional tumor size measurements in a large sample of mixed tumors, found that 52% had lost AAO-HNS class A/B hearing and 42% WRS class I/II after 4 years, which is comparable to our findings in purely intracanalicular tumors (table 6).

After 19 months' observation of a small sample of 9 nongrowing intracanalicular tumors, Warrick et al. [1999] found a loss of class A/B hearing in 25%. In their study of 74 patients with mixed tumors, Tschudi et al. [2000] reported that 33% lost hearing within 0–30 dB PTA and 35% lost hearing within 0–50 dB PTA after 35 months. We found an overall loss of class A/B hearing in 53% of the 156 patients, who were followed for an average of 55 months (table 2). The numbers are well in accordance with each other, considering the progressive nature of the hearing loss.

In 26 patients primarily operated for a purely intracanalicular vestibular schwannoma, Nadol et al. [1996] found a correlation between largest tumor diameter and pure tone loss for the low frequencies, but not for the PTA. The latter is confirmed in the present material and the former could be related to the greater loss at low pure tone frequencies in growing tumors found by Stangerup et al. [2006a].

Classification of Hearing

Several grading systems have been proposed to evaluate the hearing capabilities of patients with vestibular schwannoma [Gardner and Robertson, 1988; Kanzaki et al., 2003]. The most generally accepted has been that of the AAO-HNS, which is based on both PTA and SDS [American Academy of Otolaryngology – Head and Neck Surgery, 1995]. According to this classification, class A and class B are considered serviceable hearing, whereas class C and class D are not (fig. 2). For some patients, however, a class C hearing may be very useful, since an ear with a discrimination score of more than 70% may be easily fitted with a hearing aid. Thus, the WRS classification based only on the speech discrimination (monosyllabic word recognition) would be more reasonable [Meyer et al., 2006], as this is the parameter determining the feasibility of providing the patient with a hearing aid (fig. 2).

Patients with Normal Speech Discrimination at Diagnosis

In the largest study of hearing in observed vestibular schwannoma patients (623 patients), Stangerup et al. [2006b] proposed the adaptation of a new category in the WRS system. In the material of mixed intra- and extrameatal tumors, it was found that patients with 100% SDS at diagnosis kept a surprisingly good discrimination throughout a mean observation period of 4 years. Eighty-nine percent remained within WRS class I, compared to 43% for patients with only a small (1–10%) loss of SDS at

diagnosis. These findings led to the proposal that patients with 100% SDS at diagnosis should be considered as a special group (WRS class 0), with an especially good spontaneous hearing preservation when observed for years. Stangerup et al. [2006b] did not specify findings on purely intracanalicular tumors. We found that the risk of loosing hearing is indeed significantly smaller in these tumors when the SDS is 100% at diagnosis, which supports the findings and proposal of Stangerup et al. [2006b].

Treatment Strategy

When assessing the hearing status of a vestibular schwannoma patient, the function of the contralateral ear is of great importance. In case of normal hearing in the contralateral ear, the hearing in a tumor ear with a discrimination score of less than 70% may not be considered useful. In case of a deaf contralateral ear, however, a tumor ear with a 50% discrimination score will definitely be considered useful. None of the present 156 patients had a combination of good hearing in the vestibular schwannoma ear and unusable hearing in the contralateral ear.

Of the 50% of the patients diagnosed with a class I WRS or a class A AAO-HNS hearing, and thus being eligible for realistic hearing preservation treatment [Sanna et al., 2004], around 50% lose their hearing class during 4.6 years of observation (fig. 4, tables 2 and 3). Reported hearing preservation rates for surgery and radiotherapy have been mainly short-term and vary tremendously, between 20 and 85% for surgery [Gardner and Robertson, 1988; Weber and Gantz, 1996; Hecht et al., 1997; Slattery et al., 1997; Staecker et al., 2000; Gjuric et al., 2001; Betchen et al., 2005; Lin et al., 2005; Arts et al., 2006; Meyer et al., 2006] and between 7 and 94% for radiotherapy [Iwai et al., 2003; Lin et al., 2005; Paek et al., 2005; Combs et al., 2005], depending highly on the criteria set for the term 'preserved hearing'. For both treatment modalities, the average hearing preservation rate between these studies is approximately 50–55%. The study by Lin et al. [2005] compared the hearing preservation during observation with that of surgery (retrosigmoid) and radiotherapy (hyperfractionated stereotactic) in a material of both intra- and extracanalicular tumors. After a follow-up of 4, 9.5 and 6.8 years, as many as 88% had lost serviceable hearing (Gardner-Robertson I/II) in the radiotherapy group (42 patients), compared to 84% in the retrosigmoid surgery group (113 patients) and 57% in the observed group (86 patients), respectively. An overall interpretation of the present and the previously published data on hearing preservation can only lead to the conclusion that an observational strategy preserves hearing approximately as good as surgery or radiotherapy, within a timeframe of 4–9 years. Thus, it can be argued that active treatment of intracanalicular vestibular schwannomas with a good hearing in both ears (SDS >70%) should await documented growth by repeated MRI, although some hearing is likely to be lost during the time between scans. As around 19% of these tumors grow into the cerebellopontine angle [Caye-Thomasen et al., 2006] and 19% of these have a class A hearing, less than 4% of all patients diagnosed with an intracanalicular tumor will be candidates for hearing preservation treatment following documented growth, when the AAO-HNS classification system is applied. Using the WRS system, the corresponding numbers are 19% with growth, of 50% with WRS class I, leaving 10% as candidates.

More data on long-term follow-up of actively treated patients are needed in order to properly compare the presently reported spontaneous risk, rate and degree of hearing loss to active surgical or radiotherapeutical treatment. However, if it is assumed that active treatment does preserve long-term hearing better than observation, the present and previous findings propose that patients with good hearing in the nontumor ear and an SDS in the tumor ear between 70 and 99% should be considered as eligible for primary hearing preservation treatment. As indicated above, patients with 100% SDS on the tumor ear are especially suited for observation, as the risk of loosing significant hearing spontaneously is very small in this group of patients. In case of a poor hearing in the nontumor ear (SDS <50%), patients with an SDS between 50 and 99% are suitable for treatment. Based on these criteria, 33% of the present patients were eligible for hearing preservation treatment at diagnosis. The treatment should be scheduled as soon as possible as the hearing loss primarily progresses within the first years after diagnosis.

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