Otology

Hearing preservation surgery in acoustic neuroma. Slow progress and new strategies

Chirurgia di preservazione dell'udito. Lento progresso e nuove strategie

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SUMMARY

Quality and rate of preserved hearing are crucial to make hearing preservation surgery a viable treatment. A long-term experience with hearing preservation surgery, with tumour size and hearing as admission criteria, was evaluated to assess which size and hearing allowed a high rate of success. The hearing outcome in relation to size of tumour and pre-operative hearing was retrospectively reviewed in a consecutive series of 115 cases of sporadic acoustic neuroma which were operated on with hearing preservation surgery. Inclusion criteria were hearing with ≤ 30 dB pure tone average and ≥ 70% Speech Discrimination Score. The size was ≤ 15 mm in the first series of 51 cases, and ≤ 10 mm in the second series of 64 cases. Pre-operative and post-operative pure tone average were measured at 0.5 to 4.0 KHz, and speech discrimination score at ≤ 40 dB above perception. Post-operative hearing within 30 dB pure tone average and 70% speech discrimination score was considered socially useful hearing and successful outcome. The change to 40 dB pure tone average and 60% speech discrimination score from a pre-operative 30 pure tone average/70% speech discrimination score was considered an acceptable outcome. Patients with a tumour of ≤ 10 mm size in the cerebello-pontine-angle and hearing within 20 dB pure tone average/80% speech discrimination score had a success rate of 76%. Patients with hearing between the 20 dB pure tone average/80% speech discrimination score and 30 dB pure tone average/70% speech discrimination score had a success rate of 41%, which increased to 53% if the limit to success was set at 40 dB pure tone average/60% speech discrimination score. Patients with a tumour larger than 10 mm or hearing worse than 30 dB pure tone average/70% speech discrimination score had a poor preservation rate. In conclusion, hearing preservation surgery on a ≤ 10 mm acoustic neuroma with good hearing had a high rate of success and appeared to be a realistic treatment option which could be integrated with observation and radiotherapy in updated guidelines of treatment.

KEY WORDS: Acoustic neuroma • Vestibular schwannoma • Hearing preservation surgery

RIASSUNTO

La qualità ed il tasso di udito conservato sono cruciali nel rendere la chirurgia di conservazione dell'udito una terapia valida. La nostra esperienza con detta chirurgia ha preso come criterio di ammissione la dimensione del tumore ed il livello dell'udito e viene studiata con il proposito di accertare quale dimensione e quale udito hanno permesso un elevato tasso di successo. Si è compiuta una analisi retrospettiva del risultato uditivo in rapporto a dimensione tumorale e udito preoperatorio in una serie di 115 casi di neurinoma acustico sporadico, operati con chirurgia di conservazione dell'udito. I criteri di ammissione sono stati un udito preoperatorio di almeno 30 dB di Pure Tone Audiometry (PTA) e 70% di Speech Discrimination Score (SDS). La dimensione era non superiore a 15 mm in una prima serie di 51 casi, e non più di 10 mm in una seconda serie di 64 casi. L'udito è stato misurato con la media tonale pre- e postoperatoria alle frequenze da 0,5 a 4 kHz e con il tasso di discriminazione vocale (SDS) a non più di 40 dB di amplificazione sopra la soglia di percezione. Un udito postoperatorio entro i 30 dB di PTA ed il 70% di SDS è stato considerato udito socialmente utile ed un successo. Un cambio a 40 dB PTA e 60% SDS è stato considerato un risultato accettabile se l'udito preoperatorio era 30 dB PTA e 70% SDS. I risultati sono stati i seguenti: i casi con tumore non superiore ai 10 mm in angolo ponto cerebellare e con udito entro i 20 dB PTA e 80% SDS hanno presentato un tasso del 76% di successo. I casi con udito preoperatorio compreso tra 20 dB PTA-80% SDS e 30 dB PTA-70% SDS hanno presentato un tasso di successo del 41%, che si eleva al 53% se il limite del successo è portato a 40 dB PTA e 60% SDS. I casi con tumore più grande di 10 mm e/o con udito peggiore di 30 dB PTA-70% SDS hanno avuto un tasso di conservazione nettamente inferiore. In conclusione, la chirurgia di conservazione dell'udito in tumori piccoli, ovvero non superiori a 10 mm e con udito buono può essere considerata una terapia valida che può essere integrata con l'osservazione e la radioterapia in aggiornate linee guida di trattamento.

PAROLE CHIAVE: *Neurinoma dell'acustico • Schwannoma vestibolare • Chirurgia di conservazione dell'udito*

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Introduction

Hearing preservation was the last goal that surgery of acoustic neuroma added to the preservation of vital functions and facial nerve. Since the historical papers by Elliot and Mc Kinney [1], House [2], Rand and Kurze [3] and the ensuing reports both with the middle cranial fossa approach [4-7] and the sub-occipital approach [8-12] hearing preservation surgery continued to progress [13 14].

Other treatment modalities, e.g. radiotherapy and observation, are challenging surgery and claim a high rate of preservation with minimally invasive treatment [15] or suggest observational treatment in small non-growing tumours [16].

The present report focuses on personal experience with hearing preservation surgery (HPS) to emphasize that this option should represent the first choice approach in small tumours with good hearing and should be integrated with observation, radiotherapy, and radical surgery in updated guidelines of treatment.

Material and methods

HPS was performed in 322 cases of sporadic vestibular schwannoma from 1976 to April 2009. The eligibility to surgery changed three times over the years (Table I) to become more and more restrictive with regard to tumour size and hearing level. The first series of 207 cases was the object of a previous article [17], the second series of 51 cases and the third series of 64 are reported herewith (Table II).

The trend to operate on smaller tumours with better hearing could not always involve strict admission criteria, as follows.

The eligibility criteria of the second series were: size ≤ 15 mm in the cerebello-pontine-angle (c.p.a.) and hearing with ≤ 30 dB Pure Tone Audiometry (PTA) and ≥ 70% Speech Discrimination Score (SDS). The results led to the third series criteria of ≤ 10 mm in size, ≤ 30 dB PTA and ≥ 70% SDS, and Auditory Brain Response (ABR) with only moderate changes. As we could not take a rigid standing on criteria which were under trial, we operated also on patients exceeding the size and hearing limit: namely with good hearing in a larger tumour, or bad hearing on the non-tumour ear, or because of the patient's choice. This approach was adopted in 32 out of 51 cases in the second series and in 35 out of 64 cases in the third series. Factors, in the second series, were PTA in 10 cases, size in 8 cases, both PTA and size in 11 cases, PTA and SDS in 3. The exceeding factors of the third series were PTA in 17 cases, size in 7 cases, both factors in 2 cases, ABR in 3, PTA + ABR in 2, size + ABR in 4 cases. The size of the tumour was determined by the largest diameter in the c.p.a. at the enhanced T1 weighted magnetic resonance imaging (MRI) [19]. The extension of the tumour to the fundus of the internal auditory canal, both at MRI and operation, was recorded, but it did not influence the choice of the approach. Impaction on the fundus occurred in 57 cases, with a firm adherence in 27 of these patients. In 32 cases, the gap between the tumour and the *fundus* was 1-2 mm, in 26 it was ≥ 3 mm. The size of the tumour for each series is reported in Table II.

Hearing level was assessed with PTA and SDS. The PTA was carried out at 0.5, 1, 2 and 4 kHz, the SDS at comfortable loudness or at 40 dB above perception whichever was less [18]. These data were classified according to the AAOHNS guidelines [18] and the Consensus Meeting on Systems for Reporting Results [19] hereafter called the Tokyo classification. The Tokyo grading is as follows:

- Class A is defined as an average pure tone hearing equal or better than 20 dB PTA and a speech discrimination score of at least 80%;
- Class B has the limit of 30 dB PTA/70% SDS;
- Classes C, D, E, F have a PTA of 20 dB steps and SDS of 10%, a better speech discrimination score than PTA makes the category of the outcome one class higher.

The tables on outcome show how the pre-operative cases of each class are distributed in the post-operative classes. Outcome of every class was also reported as percent rate of the post-op class or classes over the pre-op class. A suc-

Table I. Study population and eligibility criteria to HPS and hearing outcome according to AAOHNS classification.

HPS in 322 cases series I, II, III Eligibility criteria	Hearing outcome AAOHNS '95, A-B classes
I 1976-2000, 207 cases Ø ≤ 30 mm, hearing 50/50	27.5%
II 2000-2002, 51 cases Ø ≤ 15 mm, hearing 30/70	32%*
III 2002-2008, 64 cases Ø ≤ 10 mm, hearing 30/70	50%*

** Including cases OFF protocol.*

Table II. Size of tumour in II and III series.

II series of 51 cases IN eligibility criteria 19 cases	Average Ø 8.63 mm Range 3-15 mm
OFF eligibility criteria 32 cases	Average Ø 11 mm Range 1-27 mm
III series of 64 cases IN-eligibility criteria 29 cases	i.c. 18 Average Ø 7.3 mm Range 4-9 mm c.p.a. 11 Average Ø 6 mm Range 4-8 mm
OFF-eligibility criteria 35 cases	i.c. 11 Average Ø 6.8 mm Range 4-10 mm c.p.a. 24 Average Ø 11.2 mm Range 1-26 mm

i.c.: intracanal tumor
c.p.a.: cerebello-pontine-angle tumor

cess was considered hearing within 30 PTA and 70 SDS which corresponds to Class A in the AAOHNS and A and B in the Tokyo system. The failure was expressed with two figures depending on considering, or not, a failure, the drop to the next lower class.

The surgical technique was the retro-sigmoid (RS) approach with retro-labyrinthine meatotomy as already described [17], in 96 cases, and the middle cranial fossa (MCF) approach in 19 cases of intra-meatal tumours.

With the retro-sigmoid approach, the patient was placed in a lateral (park bench) position with the head contra-laterally turned of 30°. The surgical table was further rotated 20-30° during exposure of the lateral end of the acoustic meatus.

Intra-operative monitoring was performed on the VII c.n. with electromyography (EMG), and the auditory system with ABR and compound action potential (CAP) of the cochlear nerve. Facial nerve function was assessed 18 months post-operatively and classified according to House Brackmann grading. The follow-up involved an enhanced MRI at 1, 3, 6, 10 years to assess a persistent tumour. Hearing was investigated with PTA and speech audiometry at 1 and 6 months and yearly thereafter. Pre-operative ABR had an important role for admission to HPS in the third series of cases. Absence of ABR was an absolute contraindication to hearing preservation. Absence of wave III, latency of wave V superior to 7 ms and I to V interpeak latency superior to 5.5 ms were considered unfavourable factors and relative contra-indications. They were evaluated together with the size of the tumour, PTA and SDS to decide admission to HPS, mostly in borderline cases.

Results

An overall view of the complete surgical material of 322 cases is reported in Table I. The first series [17] involved the elegibility limit of 30 mm size, 50 dB PTA and 50% SDS. The complications of the second and third series are reported in Table III. The facial nerve outcome of the 115 cases is reported in Table IV. Overall, 94% of the cases had a House Brackmann (HB) degree 1 or 2, 6% HB 3. The 7 cases with HB 3 paresis were 2 intra-meatal tumours operated on with the MCF approach, 5 cases with the RS approach and size 4, 22, 22, 26, 17 mm, respectively. In the group of 19 intra-canalicular tumours, operated upon with the MCF approach, there were 4 cases of HB 2 and one case of HB 3. In the group of 54 cases of RS approach, in ≤ 10 mm tumours the HB outcome was 1 in all cases except for two cases of HB 2 and one of HB 3. In one of the 22 mm tumours, the anatomical continuity was lost and re-established at the lateral end of the meatus with a recovery to HB 3 function.

The hearing outcome of the second and the third series is recorded for the group within the eligibility criteria and the group beyond eligibility, according to the AAOHNS and Tokyo systems. In the eligible group of the second series (Table V), the success rate was AAOHNS Class A 37%, Class A+B 74%. According to the Tokyo system, the rates were 30% Class A and 60% Class A+B. The failure rate ranged from 26% to 53%, according to the pre-operative class and which post-operative class was considered a success.

The third series had AAOHNS rates of 48% post-operative Class A and a 76% Class A+B. The failures as Classes C and D were 24% but increased to 51% if Class B was considered a failure. The rates according to the Tokyo classification were post-operative 42% Class A and 74% Class A+B for the pre-operative Class A cases. In the pre-operative Class B, the outcome was 40% Class B and 70% Class B+C. The failures were 39% or 27.5% if C Class of pre-operative B was or was not considered a failure.

The cases which were beyond the admission limit because of size and/or hearing amounted to a total of 67 for the two series and had a failure rate ranging from 89% to 76% both in the AAOHNS [18] and Tokyo system [19], respectively, and success of 37% to 50% in A cases and 23% to 16% in B cases (Table VII).

A total of 42 cases of the entire case material entered the limits of 10 mm size, 30 dB PTA and 70% SDS and are grouped together in Table VIII. This group is representative of the current criteria of elegibility to HPS and forms the end-point of our experience. With the AAOHNS system [18], Class A occurred in 48%, Classes A+B in 83%. More details are seen with the Tokyo system [19]. Class A occurred in 26% and Classes A and B in 62%. The outcome may be more appropriately re-

Table III. Complications in 115 cases (II-III series).

Patients	Complications	Outcome
F, 58 y, RS a.	Cerebellar infarction → revision surgery → haemiparesis	Regression to normal
M, 49 y, RS a.	CSF leak 7 months postop → revision surgery	Normal
F, 47 y, RS a.	CSF leak, post-op → revision surgery	Normal
M, 32 y, MCF a.	CSF leak, post-op → revision surgery	Normal

y: years; a: approach; RS: retro-sigmoid; MCF: middle cranial fossa

Table IV. Facial nerve function, post-operative outcome, 115 cases (II-III series), according with House Brackmann grading.

Post-op. VII HB°	No. patients	%
I	98	85.2%
II	10 (1 case II° HB pre-op)	8.6% (I-II° HB: 93.9%)
III	7	6.2%
Total	115	

Table V. Hearing outcome in 19 eligible cases (≤ 15 mm, ≤ 30 dB PTA/70% SDS) of II series.

AAOHNS	No. cases	Post-op. A	Post-op. B	Post-op. C	Post-op. D		
Pre-op. A	19	7	7	/	5		
TOKYO	**No. cases**	**Post-op. A**	**Post-op. B**	**Post-op. C**	**Post-op. D**	**Post-op. E**	**Post-op. F**
Pre-op. A	10	3	3	1	/	/	3
Pre-op. B	9	/	3	3	1	/	2
Total	19	3	6	4	1	/	5

Table VI. Hearing outcome in 29 eligible cases (≤ 10 mm, ≤ 30 dB PTA/70% SDS) of III series.

AAOHNS	No. cases	Post-op. A	Post-op. B	Post-op. C	Post-op. D		
Pre-op. A	29	14	8	1	6		
TOKYO	**No. cases**	**Post-op. A**	**Post-op. B**	**Post-op. C**	**Post-op. D**	**Post-op. E**	**Post-op. F**
Pre-op. A	19	8	6	2	/	/	3
Pre-op. B	10	/	4	3	/	/	3
Total	29	7	9	5	/	/	6

Table VII. Hearing outcome in 67 non-eligible cases of series II and III.

AAOHNS		No. cases	Post-op. A	Post-op. B	Post-op. C	Post-op. D		
Pre-op.	A	19	7	/	2	10		
	B	39	1	8	5	25		
	C	9	/	/	1	8		
Total		67	8	8	8	43		
TOKYO		**No. cases**	**Post-op. A**	**Post-op. B**	**Post-op. C**	**Post-op. D**	**Post-op. E**	**Post-op. F**
Pre-op.	A	14	/	7	/	/	/	7
	B	30	1	3	3	1	/	22
	C	19	/	/	3	5	1	10
	D	4	/	/	/	1	/	3
Total		67	1	10	6	7	1	42

Table VIII. Hearing outcome in 42 (≤ 10 mm, ≤ 30 dB PTA/70% SDS) cases of II and III series.

AAOHNS	No. cases	Post-op.. A	Post-op. B	Post-op. C	Post-op. D		
Pre-op. A	42	20	15	1	6		
TOKYO	**No. cases**	**Post-op. A**	**Post-op. B**	**Post-op. C**	**Post-op. D**	**Post-op. E**	**Post-op. F**
Pre-op. A	25	11	8 (3)	2 (2)	1	/	3
Pre-op. B	17	/	7 (4)	6 (4)	1	/	3
Total	42	11	15 (7)	8 (6)	2	/	6

(…): cases with upgraded class.

ferred to the pre-operative Class A and Class B and details whether the post-operative hearing kept the class or dropped by 10dB/10% score to the next lower class. The Class A cases kept their Class in 44%, and dropped to the B Class in 32% cases, thus totalling 76% within the B Class or 64% within the 30 dB PTA and 70% SDS level. This discrepancy between rate of the class and rate of the PTA/SDS level is due to the class upgrading in cases with class of SDS better than class of PTA, as set by the Tokyo classification [19]. There were 7 cases upgraded to B from C Class. That is four pre-operative A and three pre-operative B. There were 6 cases upgraded from D to C, three belonging to pre-operative A, three to pre-operative B. Overall, the rate of upgrading was 31% in the selected series of 42 cases, it was 19% in the complete series of 115 cases.

If the class upgrading is not applied, as set by the Sanna classification [19], the success is less; 64% of the Class A remain within Class A or B and 53% of Class B within Class B or C.
The Class B cases had 41% within the class and a total of 76% within the C Class and 53% in the 40 dB PTA and 60% SDS limit. Overall, 76% of the 42 cases fell within the next lower class. The failures with the AAOHNS were 52.5% as classes B, C, D or 17.5% as Classes C, D. With the Tokyo class the failures were 33% or 19% if C Class was not considered a failure in pre-operative B cases. It is noteworthy to record the post-operative change of PTA. The 25 pre-operative Class A cases were 19 post-operative Class A-B cases; there was no loss in 6 cases, and an average loss of 14.6, range 5 to 30 dB, in 13 cases. The 17 pre-operative Class B cases were 13 post-operative B or C Class cases, with no loss in 2 cases and an average loss of 15 dB, range 3 to 31 dB, in the remaining 11 cases. Recording of the post-operative PTA adds information to the class grading.

Discussion

Hearing outcome

Unilateral profound hearing loss or deafness are a common evolution both in untreated or treated acoustic neuroma and involve such disabilities as the understanding of speech in noise, localizing a sound source [20-22] and an unpleasant feeling in a noisy context. Rehabilitation of the deaf ear with the contra-lateral routing of signal or with the bone anchored hearing aid [23] eliminate the head shadow effect but do not restore the binaural hearing [24].
Hearing preservation of the tumour ear is the natural goal of every treatment. HPS over the years faced a series of unfavourable conditions. They were: the limited diffusion of this treatment, the rarity of cases, the difficulty of obtaining good results and the widespread acceptance of non-surgical treatments.
Our 30 years' involvement with HPS passed through different stages with different indications and outcomes (Table I). Outcomes of the first series of 207 cases [17] showed that only the few small tumours with good hearing allowed a reasonable rate of success. For example, 45 out of 68 cases of preserved hearing, in the group of 150 cases, had a ≤ 10 mm size, 13 had a 10-15 mm size and 10 were over 15 mm. The admission criteria were then changed in the second and third series with the aim of improving rate and quality of preserved hearing. There followed an improvement in hearing preservation (Tables V-VI) which took place with the more restrictive admission criteria up to the conclusive limit of 30 dB PTA, 70% SDS and 10 mm size. Tumours beyond this limit showed a substantial drop in outcome quality (Table VII).
Although a satisfactory hearing level cannot be judged only on the basis of audiometric criteria for the worse hearing ear [18], the hearing outcome is currently being considered a success if the level is equal to, or better than, 30 dB PTA and 70% SDS [25], which corresponds to the AAOHNS Class A and to the Tokyo Classes A and B. Tables V and VI report the number of Classes A and A+B for the two systems. The Tokyo Class B includes the cases with a PTA Class C and SDS Class B or A, which are promoted to the next higher class than PTA. The last series (Table VI) with a success rate of 48% to 76% of the AAOHNS [18] and 42% to 74% of Tokyo [19] reflects the improvement with respect to the previous series. The conclusive point of our experience is outlined in Table VIII which groups together the 42 cases of 10 mm size belonging to the second and third series. The Tokyo Classification yields a success rate as shown in Tabella VIII. Tumours with a pre-operative hearing equal to or better than the 20 dB PTA and 80% SDS remain in the B Class at a rate of 76% and within the 30/70 level in 64%. The cases with pre-operative hearing ranging between the 20/80 and 30/70 level keep their Class B in 41% and drop into class C in 35%, totalling 66% within the next lower class and 53% within the limit of 40 PTA and 60 SDS. The slight loss produced by surgery then allowed success with the 20 dB PTA/ 80% SDS cases, whereas it pushed the 30 dB PTA/70% SDS cases to the 40 dB PTA/ 60% SDS level. Whether this Class C may be a true failure, with no benefit to the patient, is not clear and also depends upon the contra-lateral ear, the binaural hearing in silence and noise and the effect of hearing rehabilitation. We conclude that HPS has a high rate of success and can be considered a realistic form of treatment in tumours size ≤ 10 mm in the c.p.a. and hearing equal to, or better than, 20 dB PTA and 80% SDS. If the hearing limit is placed at 30 dB PTA/70 SDS, the indication to HPS is still advisable.
The class upgrading, when SDS belongs to a better class than PTA, as set by the Tokyo classification, disrupts the equivalence B Class 30 dB PTA/70 SDS, or C Class 40 dB PTA/60 SDS. Adopting the AAOHNS [18] or the modified Sanna [19] classifications, which do not use the class upgrade, results in the PTA alone dictating the class. This occurs as, with diverging PTA and SDS, the worst is selected, and PTA is constantly worse, at least in our material.

Table IX. Guidelines of treatment in Acoustic Neuroma (Non-cystic acoustic neuroma, size as largest diameter in c.p.a.).

Size	Hearing	Treatment
≤ 10 mm	Good hearing Bad hearing	→ HPS → * Observation
10 to 15 mm	(Good/ Bad hearing) MRI 1 year	Growth → S or RT No growth → Observation
15 to 25 mm		→ Surgery or RT
> 25 mm		→ Surgery

** Observation involves MRI follow-up after 1, 2, 3, 4, 5, 7, 9,14, 20 years. Age, co-morbidity, surgical risk can address to non-surgical treatment.*

The small size and good hearing were commonly considered favourable prognostic factors to HPS [26]. The good results in this group of patients, however, were not obvious in the large series including cases with a lesser pre-operative hearing. Disregard of this aspect was present in critical reviews on HPS [25] and in a cautious attitude on the role of HPS [16]. These judgements contributed to the acceptance of the non-surgical treatments which therefore prevented the patients from being offered a hearing preservation option.
The dilation of the internal auditory canal caused by the tumour was demonstrated to be of prognostic value. When the diameter of the canal, as seen at bone window CT was much larger than the normal side, the cochlear nerve was thinned out by the tumour and more likely to loose function at dissection. The monitoring of the CAP (compound action potential) allowed a brief interval of time between the surgical action and its effect on the cochlear nerve function. The noxious manoeuvre could be, to some degree, corrected. The nerve function usually changed or was lost at dissecting between the posterior pole of the tumour and the meatus. It seemed that a direct, or even an indirect, distension of the cochlear nerve was the most common factor to damage it or make it vulnerable to a further, even slight, handling.
In the literature, extension of the tumour to the fundus of the auditory canal was felt to contraindicate the retrosigmoid approach, as it implied a blind dissection with risk of damaging the nerves or leaving a residual tumour. The retro-labyrinthine meatotomy, as well as the endoscopy-assisted approach [27] can solve this problem. We had no residual tumour [28] or nerve losses, at dissection, on the distal half of the canal.
Understanding the differences between classifications is important for their convenient use. The PTA at 0.5 to 3 KHz is likely to be better than a PTA at 0.5 to 4 KHz and to put a borderline case in a better class. Testing the SDS with a limited amplification, for example at 40 dB above the threshold [18] involves often a lower figure than would be without a limit. We adopted this more realistic method also with the Tokyo classification [19]. The Word Recognition Score [29] gave a 10% higher figure of success than the other systems in our material. The Tokyo A and B classes outlined better the top quality outcome and demonstrated better the correlation between a good pre-operative hearing and success. Stratifying the hearing levels in classes may involve an unnoticed effect. Hearing may change to a lower level and still keep the original class, or it may be subject to a small change and drop to the lower class. This fact is likely to take place with the hearing levels situated at the two extremes of each class.

Treatment options

Experience shows that HPS is mainly directed to the small tumour for which other options such as observation and radiotherapy are available. A rational choice of treatment can be pursued on the basis of the natural course of the disease and the outcome of the active treatment. This observation is supported by a strong argument, the spontaneous and stable arrest of growth in 71.1% of tumours lying in the c.p.a. and in 83% intra-meatal tumours [16]. Stereotactic radiotherapy claims a no-growth rate of as much as 94-98% [15 30 31]. Two points deserve attention. One is how these treatments affect hearing, the other which is the outcome of the cases submitted to secondary surgery because of growth. Observation involves a loss of hearing even in non-growing tumours [32-34]. Successful radiotherapy leads to no change in hearing in the short term [15 30] but progressive and severe loss in the mid-long term is a common experience only recently reported [35-37]. The second point concerns the principle that the outcome of observation and radiotherapy should include also the results of secondary surgery on the growing tumour. The observation in an extra-meatal tumour involves surgery when the tumour shows a detectable growth > 15 mm, which takes place in 29% of cases [16]. This means surgery in tumours over 18 mm, in which the predictable outcome is deafness, and a VII cranial nerve loss of 3 HB degrees or worse in approximately 17% of cases [38].
The intra-meatal tumour has a 17% chance of growing extra-meatal [16] while the tumour remaining intra-meatal shows a spontaneous loss of class A-B hearing in approximately 50% or more of cases [33 34]. While surgery and observation have the support of reliable studies, radiation requires the support of methodologically correct studies. The current reports take the lack of growth as an index of success and point to indicate a 94-98% rate of success [15 30 31]. This is, however, an equivocal figure, as the vast majority of small tumours spontaneously stop growing. RT should, therefore, be used in the case of growing tumours. Observing a tumour in order to treat it when growth is proven, is in itself a further cause of hearing change adding up to the late effect of radiation. Which is the efficacy of RT on a small growing tumour? No reports on this subject are available yet. We can arbitrarily combine data from various sources and reach a provisional answer. If the failure rate of 2-6% is referred to the 29% of growing tumours, as the natural history states [16], the corrected failure rate should be in the range of 7-22%. The necessary operation would result in deafness, and a facial loss of at least 3 HB degrees in a high rate of cases [39 40]. In conclusion, the overall outcome of the complete course of treatment with observation or radiation in small tumours with good hearing includes a succession of losses, e.g., the initial hearing loss, the hearing loss of the observation, and the further deafness and facial loss of the secondary surgery. The amount of these sequelae is not inferior to those produced by HPS in the complete group of similar cases and may reverse the view that observation or radiation are more conservative. HPS involves, however, other and less favourable aspects to be discussed. These are the morbidity of the surgery and the long-term decay of the

preserved hearing. Our patients returned to their pre-operative life and work within one to three months, with the exception of 5 cases which took longer. The post-operative pain in the back of the neck, radiating to the head, faded away after the first year. These problems, with their loss of quality of life, should be weighed against the lifetime psychological burden of harbouring a "benign tumour". The preserved hearing is subject to long-term decay with a different rate and degree [35]. In a ten-year follow-up of our first series, this was proportional to the initial post-operative loss. The hearing of the AAOHNS Classes A and B dropped by one class with a 16% rate of cases

Updating the guidelines of treatment

The increasing number of small tumours and the new information on the natural history and the outcome of surgery and radiotherapy offer the opportunity to update the guidelines of treatment. This involves a balance between such different factors as size and growth of the tumour, hearing and facial nerve function, age and co-morbidity, desire of the patient, as well as the burden imposed on his/her physical and emotional condition. The minimal morbidity is the common goal both of active treatments and observation. Tschudi et al. [41] suggested watchful expectation and surgery or radiotherapy in the event of proven growth. Stangerup et al. [16] proposed observation for tumours smaller than 15 mm and active treatment for tumours growing > 2 mm above this size. The 15 mm limit was to avoid the increased surgical morbidity on the VII nerve with a larger tumour. This criterion, however, entails operating on ≥ 18 mm tumours with a risk of facial nerve loss which is reported to be as high as 17% of HB Grade III or worse in the 15 to 25 mm tumour [39]. Surgery on smaller tumours lowers the facial risk to 9% [38], but it requires the non-growing tumour be excluded from unnecessary treatment. To this aim, an MRI control within the next 12 months, showing absence of growth, can be a predictor of no growth [41-43] with a 5.6% chance of missing a later arrest of growth [44].

Our guidelines (Table IX) include HPS, conventional surgery, observation and radiotherapy, and select the treatment on the basis of size and growth of the tumour, hearing and the patient-related factors such as age, co-morbidity, surgical risk and the individual's preference. If a patient with a ≤ 10 mm tumour presents good hearing within 30 dB PTA/70% SDS and ABR with only moderate changes, HPS is offered as first choice treatment. The only one exception could be a tumour with 100% SDS, in which the chance of good long-term hearing [34] suggests observational treatment. If hearing is bad, observation with MRI follow-up is the best option, as it allows treatment of the non-growing tumour to be avoided. A tumour between 10 and 15 mm in size is submitted to active treatment when annual MRI shows evidence of growth. A > 15 mm tumour involves surgery or radiotherapy. The surgical option changes to observation or radiotherapy depending on patient-related factors. Age, and/or co-morbidity with increased surgical risks indicate the need for observation or radiotherapy. The patient's desire to choose between different options is decisive. The case of the > 10 mm tumour with good hearing and a lesser chance of hearing preservation is a matter for patient's and doctor's judgement.

Updating the treatment of acoustic neuroma involved the evaluation of the complete course of each option including the management failures, and indicated the treatment with the least morbidity. Timely treatment is more effective with early diagnosis especially when active treatment is debated. The suggested guidelines, indicate that radiotherapy needs better data regarding its efficacy on the growing tumour. Early surgery on a small tumour, on the other hand, can benefit more from complete data on the rate and pattern of tumour growth.

Conclusions

Unilateral hearing loss, or deafness, are common events in acoustic neuroma and cause serious handicaps to inner, social and professional life. The HPS addressed to this problem was of limited success and raised doubts of being an unrealistic goal due to the few eligible cases and insufficient rate of success. More recent progress, however, appears to favour the important role of HPS. These include early diagnosis of small tumours, the selection of cases with a favourable surgical prognosis, the advances made in surgery thanks to the experience of the surgeon and use of monitoring. This report on our experience with HPS aims to contribute to the present debate and focuses on two points. Does HPS offer a benefit which makes the operation worthwhile? Are there alternative treatments?

1. In our experience, HPS offers a success rate of 76% in small tumours with good hearing. The limit of elegibility to HPS is size ≤ 10 mm in the c.p.a. and hearing equal to or better than 20 dB PTA and 80% SDS, and ABR with only moderate changes. Cases below this limit, but still within the 30 PTA and 70% SDS, have a 66% chance of maintaining the same class or the class immediately below.
2. Alternative treatments should be judged on the basis of the outcome of the entire course of the treatment. Observation and STRT are the options allowing a non-surgical approach, with the functional preservation and maintenance of the tumour. With both these options, however, the better preservation, which has been claimed, is not obtained. Hearing is inevitably impaired and the facial nerve is subject to a higher risk if the complete course of treatment, with the obligatory surgery on a growing tumour is accounted for.

HPS presents its own drawbacks, e.g. the fate of hearing cannot be predicted in the individual case, the operation is itself a disease. These inconveniences represent the price for recovery and belong to the balance of "pro and contra" to be discussed with the patient.

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