Hearing improvement after vestibular schwannoma surgery in the era of the hearing preservation rule – case report and literature review

Zdenek Fik, Jan Lazak, Silvie Hruba, Zdenek Cada, Eduard Zverrna, Jan Betka

Objectives. Hearing preservation after vestibular schwannoma (VS) surgery remains a surgical challenge. In some patients with preserved inner ear function, hearing improvement is achievable. As it is currently impossible to determine which patients will present this outcome, predictions must rely on previously published reports. Our case report describes a patient who experienced hearing improvement from an unuseful level to a useful one after vestibular schwannoma surgery.
Methods. Surgery was performed via suboccipital retrosigmoid approach. The patient underwent a basic audiovestibular protocol before and after the surgery – pure tone and speech audiometry, otoacoustic emissions, auditory brainstem responses, electronystagmography – together with a detailed questionnaire study. Usefulness of hearing was evaluated using the AAO-HNS guidelines, supplemented by a frequency of 4 kHz.
Results. Hearing was preserved and even improved from an unuseful level to a useful one. Based on the available literature, the most informative predictive factors for such a result seem to be: sudden sensorineural hearing loss prior to surgery, elicitable otoacoustic emissions and the origin from the superior vestibular nerve.
Conclusion. There are a limited number of studies on this topic and it is still impossible to regularly improve hearing in properly selected patients. Furthermore, the importance of postoperative hearing quality compared to other symptoms and complications remains debatable.

Key words: vestibular schwannoma, hearing improvement, retrosigmoid approach, quality of life

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INTRODUCTION

The success of hearing preservation varies in the literature from 17% to 100% and depends on the size of the tumour, the choice of surgical approach and the surgeon's general attitude toward the issue[1].

The mechanism of hearing loss in patients with VS is complex. In addition to compression of the auditory nerve by the tumour, restriction of the vascular supply of the inner ear and parasitism by the tumour (steal phenomenon), the influence of tumoural paracrine activity has been proposed as another responsible factor in recent years[2].

The hearing damage caused during surgery stems from several factors which can either be positively or negatively influenced. These are direct damage of the auditory nerve by manipulation and damage of the vascular supply of the inner ear[3]. In addition, concerning vascularization of the inner ear, it is necessary to account for a typical property of cerebral vessels - the potential to vasospasm during irritation.

In general, the highest chances for hearing preservation are presented with tumours up to 15 mm in size, which can be defined as the „hearing preservation rule" (PTA <50 dB, SDS> 50%, largest dimension < 1.5 cm). This rule can be used to detect patients in whom it is desirable to try to preserve hearing[4,5].

Finally, the decision of whether to preserve hearing is seen in a completely different light when considering the possibility of hearing improvement after the surgery. A limited number of studies on this topic exist and we will present our own experience with such a result, together with possible predictive factors.

LITERATURE SEARCH AND SELECTION

A systematic search of the literature was completed using the PubMed/MEDLINE database for articles on hearing preservation in vestibular schwannoma surgery published till January December 2019, using the keywords "vestibular schwannoma," "acoustic neuroma/neurinoma," "hearing preservation," "hearing restoration," "hearing improvement,"in the syntax below, yielding 385 papers.

Pubmed Search Syntax
((“vestibular schwannoma”) OR (“acoustic neurinoma”) OR (“acoustic neurinoma”)) AND ((“hearing improvement”) OR (“hearing preservation”) OR (“hearing restoration”)) AND (surgery OR microsurgery)

Table 1. Comparison of preoperative and postoperative audiometry.

		Tone audiometry (Hz)					Speech audiometry				
		500	1000	2000	3000	PTA	SDS (%)	SRT (dB)	MD (%)	MD level (dB)	Class
Preoperative	operated	80	70	75	85	78	60	55	100	80	C
	non-operated	20	20	25	25	23	100	23	100	40	A
Postoperative	operated	30	30	30	35	31	100	27	100	40	B
	non-operated	15	20	20	25	20	100	23	100	40	A

PTA – pure tone average (dB) on 500 Hz – 3 kHz, SDS – speech discrimination score, SRT – speech reception treshold, MD – maximal discrimination, MDlevel – level of maximal discrimination.

Abstracts were screened for relevance based on predetermined inclusion criteria and exclusion criteria. Duplicates were removed.

Inclusion crietria
1) complete journal articles
2) vestibular schwannoma (acoustic neuroma) only tumor type
3) surgical treatment regardless surgical approach

Exclusion criteria
1) inadequate audiometric data
2) opinion, editorial, or review articles
3) non-English language
4) use of any kind of radiotherapy

All papers were read and their references exhaustively checked for additional papers not occurring in the original search. Papers with any result of hearing improvement after surgical therapy has been included in the study and the list of articles can be seen in the Table 2.

CASE REPORT

The 32-year-old patient has been observed at the Department of Otorhinolaryngology and Head and Neck Surgery, 1st Faculty of Medicine, Faculty Hospital Motol since 2014 for small intracanalicular VS in the left ear with a size of 9x5x3 mm, grade I according to the Koos classification (Fig. 1a). The patient had experienced 3 months of hearing loss with tinnitus and headaches around the left ear, without imbalance problems. This was the patient's reason for seeing a physician. Apart from hearing loss, further clinical examination did not show any pathology. Control magnetic resonance imaging half a year later showed that the tumour was stationary, as were the patient's symptoms. Subsequently, the patient appeared in the outpatient office 3 years later due to impaired hearing on the left side. The control MRI showed a growth progression of +2 mm to 11x7x5 mm. At this point the tumour was grade II according to the Koos classification (Fig. 1b). Audiologically, the patient presented useless hearing Class C according to AAO-HNS guidelines (PTA 78 dB measured on 500, 1000, 2000, 3000, 4000 Hz; SDS 60%; measured 5 days before the procedure) with elicitable otoacoustic emissions (DPOAE) on both sides (Fig. 2a). The thresholds for the individual frequencies of both ears and speech audiometry are shown clearly (Table 1). Electronystagmography showed no disorder. Subjectively, the patient was no longer able to take phone calls with the affected ear. Furthermore, he reported tinnitus, paroxysmal headaches and no imbalance issues. After discussing all therapeutic options, taking into account the

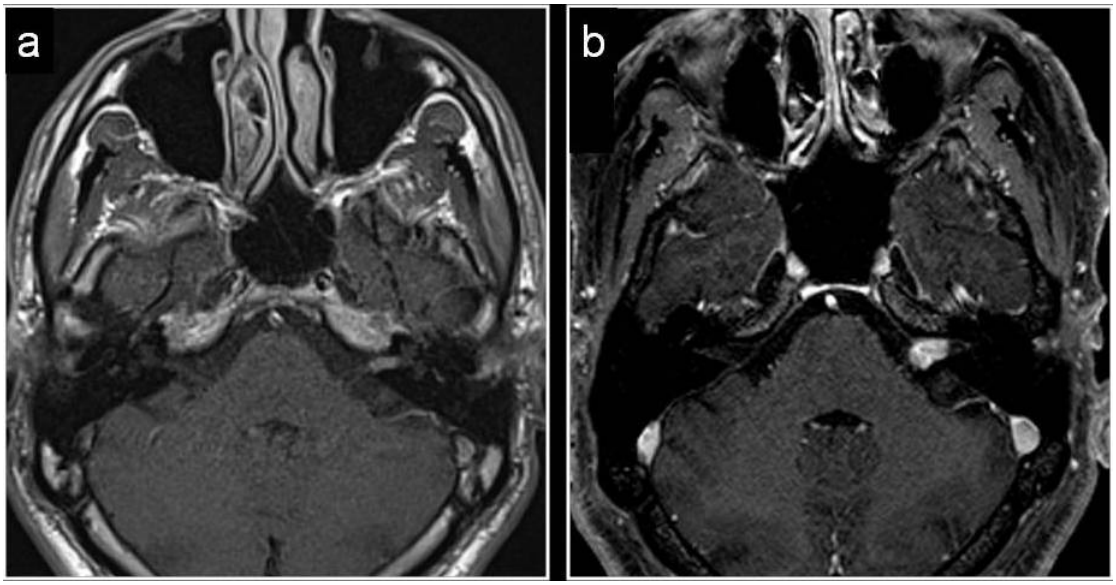

Fig. 1. Preoperative MR – progression of vestibular schwannoma on the left side: a) year 2014; b) year 2018.

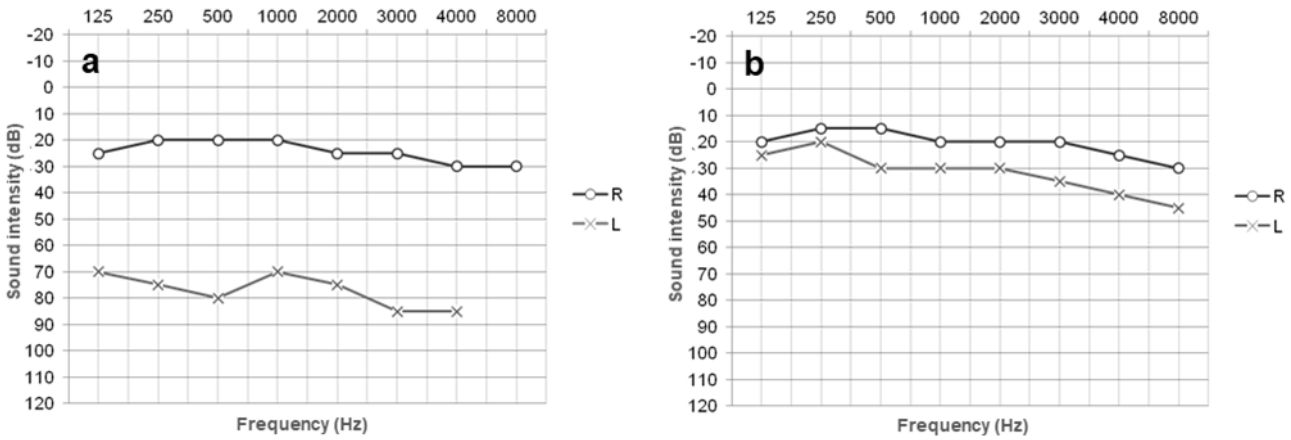

Fig. 2. Audiogram: a) 5 days prior the surgery: L – operated ear; b) 3.5 months after surgery: L – operated ear.

activity of the tumour, the patient's age and his personal preference, the surgical solution was chosen.

In 2018, the patient (36-year-old) underwent extirpation of the VS via retrosigmoid suboccipital craniotomy by opening of the internal auditory canal. Perioperative monitoring of evoked auditory potentials was not used due to preoperative useless hearing. After the exposure of the cerebellopontine angle, a small, extracanalicularly propagating tumour of typical appearance was visible. This allowed immediate identification of the facial nerve and vestibulocochlear nerve. To allow better space for preparation, we initially opened the internal acoustic meatus deep into the fundus and began to remove the tumour from the meatus. The preparation was challenging, especially in the case of the facial nerve, which ran directly ventrally from the tumour and was widespread in the area of the entrance to the internal auditory canal. The tumour was radically removed while maintaining a good stimulatory response of the facial nerve and intact auditory nerve. Both portions of the vestibular nerve were discontinued, the tumour arose from the supeior vestibular nerve (Fig. 3). The duration of the procedure from incision to suture was 5 h.

The postoperative course was uneventful, without complications. The facial nerve function has been intact from the first postoperative day (1st degree according to House-Brackmann), only a slight decrease in active motor units was described during electromyography, tinnitus disappeared, vestibular compensation was reached quickly.

Wound healing was normal. Shortly after the operation, the patient stated that he was again able to make a telephone call with his operated ear. Audiologically, one week after the procedure, the PTA on the operated ear was measured at 43 dB, speech audiometry is not routinely performed immediately after the procedure.

Three and a half months after the surgery, the average hearing threshold was 31 dB, SDS 100% (Fig. 2b, Table 1), DPOAE were elicitable. Thus, according to AAO-HNS, hearing improved to a useful level (Class B). The only dominant complaint was neuralgic headaches at the wound site.

No difference was found when comparing the examination of stem evoked potentials before and after surgery. Bilateral supra-threshold responses with latencies within the population norm, without interaural difference, were described bilaterally. No retrocochlear disorder was demonstrated.

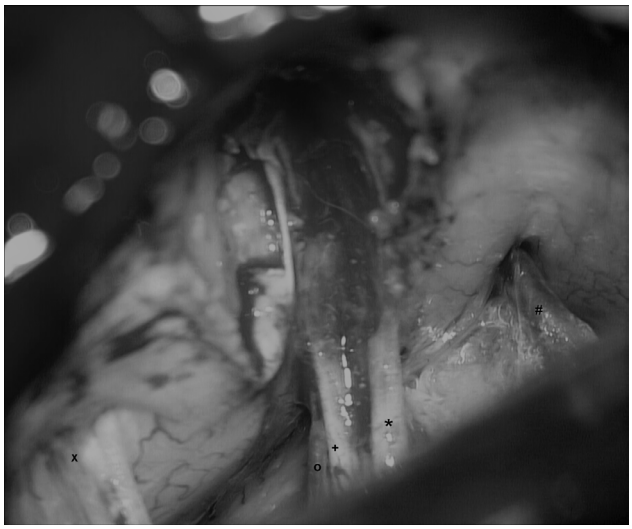

Fig. 3. Situation in the cerebellopontine angle and internal meatus at the end of the surgery: x – lower cranial nerves, o – labyrinthine artery, + – cochlear nerve, * – facial nerve, # – superior petrosal vein.

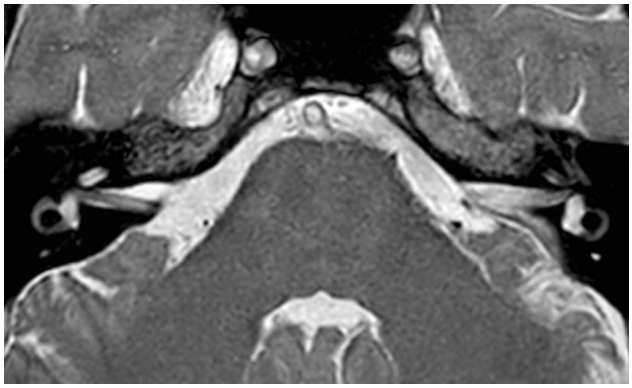

Fig. 4. Postoperative MRI.

On routinely performed magnetic resonance imaging 3 months after the surgery we saw a common postoperative condition, without signs of recurrence, with preservation of the nerve structures of the internal auditory canal (Fig. 4).

A comparison of postoperative questionnaires revealed only minimal changes in the subjective perception of hearing impairment (Hearing handicap inventory): 4/100 preoperatively versus 0/100 postoperatively. No significant difference was found before and after surgery in the other questionnaires examining the individual symptoms, with the exception of the perception of headache (Headache handicap inventory). According to the patient, there was a distinct deterioration: 36/100 vs 50/100 in this questionnaire. Bothersome headaches, however, were already present before surgery. A comparison of the questionnaires monitoring the overall condition of the patient (PANQUOL - Penn Acoustic Neuroma Quality of Life Scale + QoL36 - Quality of Life 36 questions) showed a significant deterioration of the score in the postoperative period: PANQUOL 35/130 vs 56/130; QoL 101/121 vs 80/121.

DISCUSSION

There is a dearth of literature on hearing improvement following vestibular schwannanoma surgery (Table 2). Unfortunately, there are large discrepancies in the definition of hearing improvement, as well as in the description of possible prognostic factors. It is noteworthy that the majority of the articles are from the 80s' and 90s', with the numbers falling sharply thereafter. From this it can be concluded that hearing improvement after VS surgery remains rare.

At the current level of knowledge it is not possible to reliably determine the factors that may affect the improvement of hearing after VS surgery. Generally, the crucial goal of the surgeon should be auditory nerve decompression and revascularization of the vessels to the labyrinth[2]. In addition to being a technically challenging procedure, there is still a lack of indication criteria for such an attempt.

Currently, one of the most discussed parameters which could predict hearing improvement after surgery is a short time interval between hearing loss and time of surgery. Several authors described patients who experienced sudden loss of hearing up to 3 months prior to surgery and who's hearing improved after the procedure (Table 2). In our patient, hearing deterioration occurred in the longer period before surgery and did not present signs of sudden worsening.

Nowadays, otoacoustic emissions (OEA) are being analyzed for prognostic information regarding the residual hearing capacity of patients with VS. However, only a few reviewed papers discussed OEA, as this examination method has only become widespread during the past two decades. In any case, OEA were positive in the majority of improved patients, ours included (Table 2) (ref.[6]).

Interestingly, an uncommonly discussed factor related to hearing preservation probability was lower electronystagmography (ENG) function on the affected side, meaning that tumoural involvement of the superior vestibular nerve can be a positive prognostic factor for such attempts[2,7,8]. This finding is in accordance with the presented case report, in which the tumour arose from the superior vestibular nerve, however it has not been proven on electronystagmography. Conversely, Inuoue et al. presented results of hearing improvement in patients with tumours originating from the inferior vestibular nerve[6]. Shelton and House did not find any correlation with ENG status[9].

The definition of hearing improvement is another controversial topic. Nadol et al. postulated hearing improvement as greater than 15% in SDS and no greater than 15 dB in SRT (ref.[14]). Shelton and House presented stricter criteria for hearing improvement after VS resection – with improvement being greater than 20% in SDS and/or greater than 15 dB in SRT (ref.[9]). We believe the latter definition better represents the real impact on the patient. Furthermore, SRT is sometimes used equivalently to PTA, however, PTA is favored nowadays. Of note, it is believed that the contrariety between PTA and SRT is useful in the detection of malingerers[24]. In our case, there is a discrepancy in preoperative SRT and PTA, however this still fits with the observed improvement from Class C to B for both variables.

Another discrepancy can be found in PTA counting, because a 3kHz measurement (recommended by AAO-HNS) is not always performed in published reports[24]. In our audiological assessments we count PTA starting at five frequencies, adding 4 kHz to produce more significant results. We note that it is possible to detect patients in the papers discussed who met the criteria established by Shelton and House (Table 2).

The question remains of whether it will be possible to achieve these results routinely using currently available equipment and pharmacological support, as the auditory nerve is much more prone to damage if manipulated when compared to the facial nerve[27]. Useful technical notes of cochlear nerve preservation have been described in the pioneering work „Hearing preservation age" by Jannetta et al.[28].

We must also not forget the negative effects of postoperative hearing preservation. Several reports mention a higher risk of persistent tinnitus, especially if the tinnitus is perceived by the patient as bothersome[29]. We also know that in an effort to preserve the auditory nerve and the delicate structures of the inner ear, the surgeon is sometimes pushed towards a less radical tumour removal, especially in the fundus of the internal auditory canal, and patients are thus more exposed to the risk of a small tumour residue remaining behind[30]. In contrast, non-radical operation can be the surgeon's strategy when attempting to preserve cranial nerve function during vestibular schwannoma surgery[16,26].

Last but not least, the question remains as to what role hearing preservation plays in the patient's overall perception of quality of life after surgery. Quality of life is slowly being included in management decisions in some centers[31]. Although no conclusions can be drawn from a single-patient questionnaire study, it can be predicted that hearing preservation will not be a priority for some patients[32]. Therefore, selection of such patients can be important in the choice of treatment strategy, both for one

Table 2. Overview of articles reporting hearing improvement results.

Author	Number of patients with improved hearing	Number of operated patients	Approach	Tumour size (mm)	Koos classifiaction	Number of patients with preoperatively unuseful hearing	Number of patients with postoperatively unuseful hearing	Improvement from unuseful to useful hearing	Improvement in SDS more than 20%	Improvement in SRT more than 15 dB	Improvement in PTA more than 10 dB	SSNHL/hearing loss up to 3 months prior surgery	Lowered vestibular function on ENG	Preoperative OEA	Preoperative ABR	Ref.
Fischer et al. 1980	1	NA	RS	30 (CT)	IV	1	1	1	1	NA	1	0	NA	not used	not used	11
Cohen et al. 1981	1	NA	RS	< 20	II	1	1	1	1	NA	1	0	0	not used	not elicited	12
Sterkers et al. 1984	5	135	RS	10 - 20	I - II	5	3	3	4	NA	3	NA	NA	not used	NA	13
Abramson et al. 1985	1	20	RS	8	I	1	1	1	1	0	0	NA	NA	not used	elicited	14
Nadol et al. 1987	1	69	RS	5	I	NA	NA	NA	1	1	NA	NA	NA	not used	NA	15
Telian et al. 1988	1	NA	RS	10 (CT)	II	1	1	1	1	NA	1	1	NA	not used	only wave I	16
Shelton et al. 1990	9	104	MFA	4 - 20	NA	NA	NA	NA	8	3	4	3	0	not used	2 patients - delayed wave V	10
Kawaguchi et al. 1994	1	NA	RS	40 (CT)	IV	1	1	1	1	NA	1	0	NA	not used	elicited	17
Yanaghira et al. 1994	3	NA	MFA	9 - 15	I - II	2	3	2	0	NA	3	3	NA	not used	3/3 elicited	18
Weber et al. 1996	2	49	MFA	NA	NA	NA	NA	NA	2	2	NA	NA	NA	not used	not used	19
Matthies et al. 1997	4	420	RS	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	not used	used	20
Ishikawa et al. 1998	2	43	MFA	NA	NA	2	2	0	1	NA	1	2	NA	not used	NA	21
Fahlbusch et al. 1998	1	61	RS	35	IV	1	1	1	1	NA	1	NA	NA	not used	only wave I	22
Noguchi et al. 1999	3	44	MFA	NA	NA	8	18	1	2	NA	1	NA	NA	not used	NA	23
Roberson et al. 1999	4	22	MFA	8 - 18	I - II	2	4	2	3	NA	1	NA	NA	not used	4/4 not elicited	24
Stidham et al. 2001	7	30	MFA	NA	NA	4	6	3	5	NA	1	NA	1	not used	7/7 not elicited or abnormity	3
Meiteles et al. 2002	1	NA	RS	10	II	1	1	1	1	1	1	1	NVS	not used	not elicited	8
Inoue et al. 2003	1	NA	MFA	NA	II	1	1	1	1	NA	1	1	NVI	elicited	only wave I	7
Arts et al. 2006	1	73	MFA	NA	NA	1	1	0	NA	NA	NA	NA	NA	NA	NA	25
Kohno et al. 2015	9	NA	RS	NA	I - IV	8	9	8	8	NA	6	9	NA	8/9 elicited	9/9 elicited	26
Sass et al. 2019	4	31	RL	NA	NA	9	5	4	2	NA	0	NA	NA	NA	NA	27

CT – computer tomography, RS – retrosigmoid suboccipital approach, MFA – middle fossa approach, RL – retrolabyrinthine approach, NA – not available, NVI – inferior vestibular nerve, NVS – superior vestibular nerve.

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of the three basic treatment modalities and for the choice of surgical approach.

From this perspective, the experience, preferences and overall philosophy towards VS surgery at individual departments come to the fore.

CONCLUSION

The presented case of VS surgery by retrosigmoid suboccipital craniotomy describes the improvement of hearing from useless to useful levels. It somewhat justifies the effort to preserve hearing in selected patients with useless hearing, good cochlear function and small tumour size despite the widespread hearing preservation rule. Sudden sensorineural hearing loss prior to surgery, elicitable otoacoustic emissions and the origin from the superior vestibular nerve (proven on ENG) seem to be useful guides for predicting these results. However, the importance of postoperative hearing quality compared to other symptoms and complications remains debatable.

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