





CLINICAL EXPERIENCE

A novel scoring system based on small vestibular schwannomas to determine consideration for cochlear implantation

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1 | INTRODUCTION

Sporadic vestibular schwannomas (VS) can be treated using radiation therapy, microsurgical resection or a wait-and-scan approach. In general, small VS are usually followed (wait-and-scan) until a significant amount of growth per year is seen.

In a recent study, 212 cases of VS were observed over time; 66% showed growth, of which 30% were fast growing (doubling in size in a year).¹ Frischer et al examined cases of VS after radiosurgery (Gamma Knife), and found a tumour control rate of 92% after 5 years of follow-up.² However, among patients with initial Gardner-Robertson class 1 or 2 hearing, only 55% maintained serviceable hearing (class 1 or 2) after 2 years, and 34% after 10 years. Several other studies have reported similar results.^{3,4} A recent meta-analysis concluded that among patients who undergo microsurgical treatment with a hearing-preserving technique, 35–49% still have serviceable hearing five years later.⁵ The above-mentioned findings support the conclusion that most VS will require treatment after some period of observation. However, the currently available treatment options carry a high risk of loss of serviceable hearing.

Some authors believe that the possibility of cochlear implantation in the setting of VS should completely change the treatment strategy for this disease.⁶ However, patient outcomes with cochlear implants (CI) show a wide variation—with some patients having no auditory perception, while others exhibit open-set speech understanding.⁷

The intraoperative testing of cochlear nerve function using electrically evoked brainstem response audiometry (eABR) provides a means of objectively assessing cochlear nerve conduction, and has been increasingly applied in this setting.^{7,8} In the present study, we aimed to assess outcomes of patients undergoing VS resection and CI, and to develop a new scoring system to preoperatively identify suitable patients for this treatment course. Appropriate preoperative classification of patients can have important impacts on patient counseling and expectations.

2 | DESIGN

Each included patient gave their informed consent to participate in the study. The trial was approved by the local ethics committee 1111/2017, and was registered at www.clinicaltrials.gov (NCT03745560).

All 17 patients underwent translabyrinthine VS resection, during which eABR were recorded with an intracochlear test electrode (ITE) before and after tumour resection. An ITE is similar to a CI electrode, but shorter and has three electrodes as well as an additional separate reference electrode. In cases with positive responses after complete tumour resection, a CI was placed. Overall, ten patients received a CI with a Flex 28 or Flex Standard electrode (MED-EL, Innsbruck, Austria) depending on cochlear duct length.

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3 | SETTING

The study was conducted at a tertiary care center.

4 | PARTICIPANTS

Seventeen patients with unilateral sporadic VS were screened between January 2017 and January 2020 (Table 1). Inclusion criteria were as follows: sporadic VS showing growth on repeat imaging studies, no ipsilateral functional hearing, desire to undergo cochlear implantation and general good health. Exclusion criteria were prior treatment for VS, neurofibromatosis type 2, history of pathology requiring multiple head magnetic resonance imaging (MRI) or history of malignant disease of the head and neck. A lack of functional hearing was defined as $\leq 50\%$ monosyllable recognition at 80 dB HL. Pure tone audiometry was performed using headphones. To reduce cross-hearing, appropriate masking was used. The appropriate masking level was determined by the experienced audiologist.

Word recognition scores (WRS) were assessed using the Freiburg monosyllables word test which is an open set German word recognition test using lists of 20 words recorded by a male talker. To minimise interaural crosstalk effects masking noise was applied to the contralateral ear, except for subjects who suffered from severe contralateral conductive HL. Here, the contralateral ear was covered with a circumaural 3 M Peltor H540A earmuff (35 dB average attenuation). WRS were measured at 65 dB SPL and 80dB SPL with loudspeakers placed at a distance of 1.5 m perpendicular to the ear being tested.

5 | MAIN OUTCOME MEASURES

The patients' preoperative findings and results were re-evaluated to develop a new scoring system for patient selection. All patients underwent MRI of the brain performed using a 3.0 Tesla magnetic resonance (MR) unit (Philips Achieva; Philips Medical Systems, Best, the Netherlands). To visualise the cerebellopontine angle, the MR protocol included a 3D balanced fast field echo sequence that was used for further assessment. Using a picture archiving and communication system (IMPAX; AGFA HealthCare, Bonn, Germany), all MR examinations were anonymised and randomly presented to a neuroradiologist who was not aware of any clinical data. Grading according to the four-grade Koos classification system was carried out.⁹ Additionally, a four-level grading system was used to evaluate intrameatal extension towards the modiolus (Table 2).

Preoperatively promontory stimulation eABR was performed in local anesthesia approximately two weeks before surgery with a gold coated rigid electrode with a rounded-bent (hockey stick) tip stimulation probe, which was placed on the promontory. The hockey stick electrode was manufactured by MED-EL (Innsbruck,

Key points

- Simultaneous translabyrinthine tumour resection and cochlear implantation is a promising treatment method with hearing rehabilitation for sporadic vestibular schwannomas.
- Size of vestibular schwannoma, distance to modiolus, residual hearing and promontory stimulation electrical brainstem response audiometry are important preoperative predictive factors for determining a positive post-tumour resection electrically evoked auditory brain stem response.
- The presence of an electrically evoked auditory brain stem response with an intracochlear test electrode after tumour resection was used to determine whether a patient received a cochlear implant.
- Positive electrically evoked auditory brain stem responses measured with an intracochlear test electrode after tumour resection seem to correlate well with auditory perception with a cochlear implant.

Austria). EABR was recorded with the Neuropack, Nihon Kohden, Tokyo. A recording window of 10 ms was used. In order to minimise artefacts, an adopted approach was used as described in the manuscript by Polak et al.¹⁰ For the stimulation, alternating biphasic pulses with the stimulating rate of 34 pulses per second was used. Phase duration was set to 100 μ s and increased in a step of 50 μ s until a response was detected. Stimulations were performed at various intensities, depending on the clarity of the response and tolerance of the patient. A positive response was defined as a clear wave V reproducible in latency and amplitude. In cases with a wave V that could only be seen at high stimulation intensities (above the level of tolerance) or that was not reproducible an unclear response was documented. No identifiable wave V was defined as a negative response. Promontory stimulation eABR with the hockey stick electrode was carried out preoperatively. In contrast, intraoperatively eABR was carried out with an ITE through a round window approach.

6 | RESULTS

Seventeen patients underwent translabyrinthine VS resection. The final decision regarding cochlear implantation was made intraoperatively, after tumour removal, based on the results of eABR with the ITE. A total of ten patients had positive eABR results and consequently received a CI.

After six months of follow-up, ten patients were daily users of the CI. Their mean aided pure-tone average was 38 dB HL, and their mean WRS was 28% at 65 dB, and 52% at 80 dB. Nine of the ten

TABLE 1 Patient demographics

ID	Age (y)	Sex	Side	4-PTA	WRS @ 80 dB	Contralateral	4-PTA _c
1	47	f	l	77 dB	20%	SSD	6 dB
2	59	m	r	68 dB	40%	AHL	54 dB
3	55	f	l	51 dB	5%	SSD	20 dB
4	74	m	r	85 dB	0%	AHL	48 dB
5	42	f	l	68 dB	0%	SSD	8 dB
6	61	f	l	64 dB	30%	SSD	5 dB
7	69	f	r	≥100 dB	0%	≥100 dB	65 dB
8	59	m	r	60 dB	0%	SSD	21 dB
9	44	m	r	74 dB	0%	SSD	6 dB
10	55	f	r	49 dB	40%	SSD	6 dB
11	56	f	l	≥100 dB	0%	SSD	10 dB
12	60	f	r	87 dB	0%	SSD	16 dB
13	44	m	l	75 dB	0%	AHL	55 dB
14	52	f	l	≥100 dB	0%	SSD	25 dB
15	56	f	l	40 dB	25%	SSD	14 dB
16	62	f	r	64 dB	0%	SSD	20 dB
17	46	f	l	≥100 dB	0%	≥100 dB	80 dB
Total	55.4 (±8.9 SD)	m = 5 f = 12	l = 9 r = 8	74 dB (±19 SD)	9% (±15 SD)	SSD =12 AHL =3	27 dB (±23 SD)

Age is given in years (y). Gender is described as f (female) or m (male). Puretone average (4-PTA) is calculated as average decibel (dB) hearing level (HL) at the frequencies 500 Hertz (Hz), 1000 Hz, 2000 Hz and 4000 Hz. Word recognition scores (WRS) are calculated using Freiburg monosyllables at (@) 80 dB HL. Hearing of the contralateral side resulted in the diagnoses of single-sided deaf (SSD), asymmetric hearing loss (AHL) or bilateral complete hearing loss larger than 100 dB HL (≥100 dB). Contralateral 4-PTA = 4-PTAC.

TABLE 2 Scoring system

Category	Definition	Points
Koos Grading	Koos 4	0
	Koos 3	1
	Koos 2	2
	Koos 1	3
Extension	Transmodiolar extension	0
	Infiltration of modiulus	1
	Contact with modiulus	2
	No contact, no infiltration of modiulus	3
Hearing	Complete hearing loss	0
	Some residual hearing (0% monosyllables)	1
	≥1% monosyllables, any PTA	2
PS EABR	No response	0
	Unclear wave V	1
	Stable wave V	2
Total Score	Class IV	0–3
	Class III	4–5
	Class II	6–7
	Class I	8–10

Scoring system to identify patients with higher chances of nerve integrity in case of VS resection. A certain amount (0-3) of points are given in four categories. Points are added up and patients are categorised to a certain class which reflects the probability of cochlear implantation after translabyrinthine vestibular schwannoma excision. Promontory stimulation eABR (PS EABR).

patients had open-set speech understanding. The patient without open-set speech understanding had significant hearing loss on the contralateral side and a language barrier.

Facial nerve outcomes were measured using the House-Brackmann (HB) score. On the first postoperative day, four of seventeen patients had a facial nerve palsy, with HB scores of 2, 3, 5 and 5. Two of these patients exhibited complete resolution at the one-month follow-up appointment. One patient showed marginal facial weakness (HB 2) at the six-month follow-up appointment. One patient exhibited permanent facial nerve weakness with a synkinesis. Preoperative imaging of this individual showed that the VS seemed to follow the facial nerve up to the geniculate ganglion, which might explain the persistent palsy.

Table 3 presents the detailed hearing results and facial nerve outcomes for each patient. In all cases, the six-month follow-up MRI showed no sign of residual or recurrent tumour. The internal auditory canal was sufficiently visible in all cases despite CI placement.

7 | THE SCORING SYSTEM

Table 2 shows the scoring system that was developed. Besides Koos grading, results of promontory stimulation eABR were included. Our system also evaluated residual hearing, which is an important factor reflecting the state of the nerve and the cochlea before an intervention. Further, we identified relation to the modiulus as a predictive factor. Table 4 shows the scoring system applied to the presented patients.

TABLE 3 Patient outcomes

ID	Koos	CI	6-month Follow-up			VII	
	1/2/3/4	yes/no	4-PTA	WRS @ 65 dB	WRS @ 80 dB	HB - POD1	HB - 6 Mo FU
1	2	yes	35 dB	40%	65%	1	1
2	1	yes	30 dB	65%	65%	1	1
3	3	no	n.a.	n.a.	n.a.	5	3
4	3	yes	36 dB	0%	30%	1	1
5	2	no	n.a.	n.a.	n.a.	1	1
6	1	no	n.a.	n.a.	n.a.	1	1
7	2	no	n.a.	n.a.	n.a.	1	1
8	4	no	n.a.	n.a.	n.a.	5	2
9	2	yes	35 dB	0%	20%	1	1
10	2	yes	34 dB	45%	85%	1	1
11	2	no	n.a.	n.a.	n.a.	1	1
12	2	yes	35 dB	10%	45%	2	1
13	2	yes	51 dB	n.p.	n.p.	1	1
14	1	no	n.a.	n.a.	n.a.	3	1
15	2	yes	33 dB	40%	60%	1	1
16	1	yes	43 dB	20%	40%	1	1
17	2	yes	35 dB	35%	55%	1	1

Outcomes of seventeen included patients. The second column shows the size and extension of the vestibular schwannoma according to Koos grading one to four. The third column shows which patients were provided with a cochlear implant (CI) (yes) and which not (no). Column four—Puretone average (4-PTA) in CI aided condition calculated as average decibel (dB) hearing level (HL) at the frequencies 500 Hertz (Hz), 1000 Hz, 2000 Hz and 4000 Hz. Column five and six—word recognition scores (WRS) in CI aided condition are calculated using Freiburg monosyllables at (@) 65 and 80 dB HL. The last two columns show facial nerve function according to House Brackmann (HB) scale 1 to 6 on postoperative day one (POD 1) and at the 6 months follow-up appointment (6 Mo FU). n.p.—not performed due to a language barrier. n.a.—not applicable.

8 | DISCUSSION

Various advancements over recent years have made CI a good option for restoring binaural hearing in patients with VS. Imaging sequences have been improved and CI magnets have been developed to reduce metal artifacts¹¹ and allow postoperative evaluation of the internal auditory canal and cerebellopontine angle for tumour follow-up.¹² Another recent development is the possible use of intraoperative eABR with an ITE, enabling objective assessment of cochlear nerve functionality.^{7,8}

In the past, the outcome of CIs in the context of VS was considered unpredictable, and many patients did not benefit from this technique. However, the advent of testing systems, such as intraoperative eABR, permits more precise evaluation of possible outcomes. Although simultaneous VS resection and cochlear implantation is possible with only visual assessment of the cochlear nerve,¹³ eABR measurements are a useful tool for objectifying the surgeon's assessment. Only two prior studies have described the use of eABR during translabyrinthine VS resection.^{7,14} One study included eight patients.¹⁴ The other was a pilot study by our research group.⁷ In this previous manuscript, we demonstrate the intraoperative objective evaluation of the cochlear nerve with eABR, using an ITE, during

translabyrinthine VS resection and cochlear implantation.⁷ Three of the patients (without any prior treatment of VS) in said previous study⁷ were included in the current manuscript. Our preliminary results indicated that positive eABR results (clear wave V) seem to be reliable, and to correlate well with CI-aided auditory perception (ie those with positive eABR results had sound perception with the CI). In contrast, eABR results have not been able to predict outcomes such as word recognition scores or speech understanding with CI. This thesis was further supported by the results of our present study, in which all patients with positive eABR results and who received an implant showed auditory perception with their CI, and most (90%) even have open-set speech understanding.

A remaining major challenge is to identify patients with a high chance of cochlear nerve preservation and positive eABR results. Therefore, as a logical next step, the knowledge gained in this study was used to create a grading system with the aim of preoperatively determining candidacy, as was performed herein.

Individuals with residual speech understanding were more likely to have positive eABR results. These results are in line with the findings of Sanna et al.¹³ In their study, individuals with good functional hearing underwent translabyrinthine VS resection and cochlear implantation. After tumour removal, surgeons evaluated the cochlear

TABLE 4 Applied point system

ID	Koos Points	Modiolus	Audio	PS eABR	Points	Class	CI
1	2	2	2	n.p.	≥6	≤II	yes
2	3	3	2	2	10	I	yes
3	1	2	2	2	7	II	no
4	1	3	1	2	7	II	yes
5	2	2	1	2	7	II	no
6	3	1	2	2	8	I	no
7	2	0	0	1	3	IV	no
8	0	1	1	2	4	III	no
9	2	3	1	2	8	I	yes
10	2	3	2	2	9	I	yes
11	2	1	0	0	3	IV	no
12	2	3	1	1	7	II	yes
13	2	3	1	2	8	I	yes
14	3	1	0	2	6	II	no
15	2	3	2	2	9	I	yes
16	3	2	1	2	8	I	yes
17	2	2	0	2	6	II	yes
Results	0–3	0–3	0–2	0–2	0–10	I–IV	yes/no

Point system applied to the presented seventeen patients. Every column represents one of the categories and points given. Koos—Koos classification, Modiolus—extension (in connection to the Modiolus), Audio, audiometric results, summation of pure tone average and word recognition score) and PS eABR, promontory stimulation electrically evoked auditory brainstem response. In total, there are four categories. Points reflects the sum of all points. Class is the resulting group each patient is categorised into, according to amount of points. Patient 1 did not undergo PS eABR, which does not allow for a complete classification. CI, cochlear implant: yes if they were provided with a CI, no if no CI was placed. N.p., not performed.

nerve and visually determined whether it was intact.¹³ Of the nineteen included patients, thirteen were able to receive a CI, of whom 84% use their CI daily or almost daily.¹³

Another identified predictive factor was the VS extension towards the cochlear modiolus. This anatomic structure also reportedly plays an important role when assessing CI candidacy in cases of malformation,¹⁵ as well as the need of its preservation in intracochlear VSs.¹⁶ Obviously, tumour resection is increasingly challenging with greater VS size, reducing the possibility of nerve protection reflected by Koos grading. The last category of our scoring system was promontory stimulation. One issue with promontory stimulation is that it produces a substantial number of artefacts in awake patients. Another problem is that some patients do not tolerate stimulation well, leading to a wide variety of stimulations intensities between different individuals. Artefacts and said stimulation range diminish the interpretation possibilities and reliability of the results.

One limitation of the present study was the relatively small sample size on which the scoring system was based. Nevertheless, this study represents the largest group of patients with VSs, in whom eABR was intraoperatively performed. As well as the first study assessing predictive factors for positive eABR results after tumour resection and possible cochlear implantation. Another important factor to keep in mind is that CI results are given 6 months after

implantation. A plateau of word recognition results is generally reached at 12 months after activation.

Overall, our results indicated that patients with a Class I scoring had a very high chance of positive eABR results after tumour resection; six of seven Class I patients received a CI. Among patients categorised as Class II (6–7 points according to our system), the majority (57%) had positive eABR results and could therefore be implanted, but they had a distinctly worse chance of favorable eABR results, and thus careful counseling is essential in these cases. Patients categorised as Class III and IV had negative eABR results and were therefore not implanted.

9 | CONCLUSION

Simultaneous translabyrinthine VS excision and cochlear implantation using intraoperative eABR measurements is a good option for hearing rehabilitation. Preoperative accurate assessment of the size and extent of VS, audiometric testing, and promontory stimulation eABR improves preoperative patient selection, helps manage patient expectations, and predicts the possibility of cochlear implantation. Future research is required to determine if cochlear implantation is possible for situations outside this scoring system or whether other

factors can determine the possibility for cochlear implantation after VS excision and how to optimise their outcomes further.

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CONFLICT OF INTEREST

CA received a research grant by MED-EL. The remaining authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

AUTHOR CONTRIBUTION

All authors contributed equally to this work. VD, CM and CA designed the study. US-N. and EY collected the data. VD, EY and AA wrote the main paper and designed figures. US-N., C.M and CA provided critical revision. All authors discussed the results and implications and commented on the manuscript at all stages.

Data availability statement

The data that support the findings of this study are available from the corresponding author upon request.

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