Otology & Neurotology
39:365–371

Cochlear Implantation After Partial or Subtotal Cochleoectomy for Intracochlear Schwannoma Removal—A Technical Report

*Stefan K. Plontke, †Sabrina Kösling, and *Torsten Rahne

**Department of Otorhinolaryngology, Head & Neck Surgery; and †Department of Radiology, Martin Luther University Halle-Wittenberg, University Medicine Halle, Germany*

Objective: To describe the technique for surgical tumor removal, cochlear implant (CI) electrode placement and reconstruction of the surgical defect in patients with intracochlear schwannomas.
Study Design: Retrospective case review.
Setting: Tertiary referral center.
Patients: Ten patients (five men, five women, mean age 48 ± 12 yr) with profound or severe to profound hearing loss due to intralabyrinthine schwannomas with intracochlear location.
Interventions: Surgical tumor removal through extended round window approach, partial or subtotal cochleoectomy with or without labyrinthectomy and reconstruction of the surgical defect with cartilage, perichondrium or temporal muscle fascia, and bone pâté. Eight patients received a cochlear implant in the same procedure.
Main Outcome Measures: Retrospective evaluation of clinical outcome including safety aspects (adverse events) and audiological performance at early follow up in cases of cochlear implantation.
Results: The tumor was successfully removed in all cases without macroscopic (operation microscope and endoscope) tumor remnants in the bony labyrinth apart from one case with initial transmodiolar growth. One patient needed revision surgery for labyrinthine fistula. At short-term follow up (3-month post-surgery), good hearing results with the cochlear implant were obtained in all but one patient with a word recognition score of 100% for numbers, and $64 \pm 14\%$ for monosyllables (at 65 dB SPL in quiet).
Conclusions: Surgical tumor removal and cochlear implantation is a promising treatment strategy in the management of intralabyrinthine schwannoma with intracochlear location, further extending the indication range for cochlear implantation. It is, however, of importance to observe the long-term outcome in these patients and to address challenges like follow up with magnetic resonance imaging.
Key Words: Acoustic neuroma—Cochlea—Cochlear implant—Intracochlear—Intralabyrinthine—Surgery—Vestibular schwannoma.
Otol Neurotol **39:**365–371, 2018.

Intralabyrinthine schwannomas (ILS) are a rare subgroup of benign tumors of the 8th cranial nerve. Due to their location in the inner ear, the tumors usually become symptomatic with hearing loss, and/or vertigo, and/or tinnitus, already when still very small. In some cases, they may mimic diseases like Menière's disease or idiopathic sudden sensorineural hearing loss (ISSHL), often resulting in single-sided deafness (SSD) (1–6).

Cochlear implantation has become a standard procedure for hearing rehabilitation in bilateral profound hearing loss or anacusis. There is growing evidence, however, of the benefits of cochlear implantation also in patients with SSD (7–12). Initial but limited reports on cochlear implantation in patients with intralabyrinthine schwannomas showed promising results with respect to hearing rehabilitation (13–17). Due to the clinical characteristics and the specifics of surgical accessibility of ILS, new options evolve for the surgical management of these tumors in combination with hearing rehabilitation, that differ from the management strategies of ''classical'' vestibular schwannomas.

We here describe the surgical technique for tumor removal, reconstruction of the defect, and safety aspects

Address correspondence and reprint requests to Prof. Dr. med. Stefan K. Plontke, Department of Otorhinolaryngology, Head & Neck Surgery, Martin Luther University Halle-Wittenberg, University Medicine Halle, Ernst-Grube-Str. 40, 06120 Halle (Saale), Germany; E-mail: stefan.plontke@uk-halle.de

Funding: There was no external funding received for this work.
The authors' institution (of SKP and TR) receives grant support for other collaborative research work (not related to this study) with Cochlear Ltd., Sydney, Australia, and Medel, Innsbruck, Austria. The authors SKP and TR received travel support for lectures from Cochlear, Ltd, Australia, and Medel, Innsbruck, Austria (not related to this study).
DOI: 10.1097/MAO.0000000000001696

TABLE 1. *Demographic data, surgical procedures, and pre- and postsurgical audiological data*

Nr.	Age[a]	m/f	Side R/L	Pre-op Hearing Loss (4PTA [dB HL]/ WRS_{max}) (%)	Tumor Location	Procedure	Post-op (3 mo) WRS_{65} Numbers/ Monosyllables (%)	Last Available Post-op WRS_{65} Numbers/ Monosyllables (%) (mo)
1	49	m	L	109/0	Intracochlear (basal)	Extended cochleostomy, partial resection of basal turn (Figs. 3 and 4 in (16))	100/75	100/95 (24)
2	60	f	R	>110/0	Intracochlear (basal)	Partial cochleoectomy, reconstruction with separation of the first and second cochlear turn with cartilage and CI-dummy insertion [Fig. 5 in (16))	Dummy, no CI	n/a
3	47	f	R	>110/0	Intracochlear (entire cochlea)	Subtotal cochleoectomy, reconstruction and CI-dummy insertion (19)	Dummy, no CI	n/a
4	60	f	R	>110/15	Intracochlear	Subtotal cochleoectomy, reconstruction and CI (Fig. 6 in (16))	30/0 (at first fitting)	Lost to follow-up
5	33	m	R	79/30	Intracochlear (middle and apical turn) and (initially multilocular: +IAC/CPA)[b]	Subtotal cochleoectomy, reconstruction and CI (removal of tumor in IAC/CPA via retrosigmoidal approach 5 yr before)[b]	100/50	100/70 (6)
6	36	f	R	>110/0	Intravestibulo-cochlear	Subtotal cochleoectomy + labyrinthectomy, reconstruction and CI	100/55	100/30 (6)
7	51	m	L	74/15	Intracochlear (middle and apical turn)	Subtotal cochleoectomy, reconstruction and CI	100/45	100/80 (6)
8	32	m	L	98/0	Intracochlear (basal turn)	Subtotal cochleoectomy, reconstruction and CI	100/85	100/95 (6)
9	48	f	R	>110/0	Intravestibulo-cochlear (initially transmodiolar[c])	Partial cochleoectomy and tumor removal by ''pull-through'' + labyrinthectomy, reconstruction and CI (removal of IAC via retrosigmoidal approach 3 yr before)[c]	100/65	100/65 (6)
10	67	m	R	80/0	Intracochlear (basal and partial middle turn)	Subtotal cochleoectomy, reconstruction and CI	100/75	100/75 (3)
	48 ± 12	5f/5m	7R/3L	$99 \pm 14/6 \pm 9$			$100/64 \pm 14$[d]	

[a]Age: age at surgery.

[b]Patient 5: On retrospective evaluation of initial MRI, a clear distinction between the vestibular schwannoma in the lateral portion of the IAC and the CPA and a (at that time undiagnosed) small ILS in the apical turn of the cochlea could be seen.

[c]Patient 9: This tumor was most likely initially a (non-diagnosed) transmodiolar tumor three years before partial cochleoectomy, labyrinthectomy, and CI the patient underwent surgical removal of an intrameatal vestibular schwannoma through a retrosigmoidal approach in the department of neurosurgery.

[d]Without patient 4.

4PTA indicates average air conducted pure tone threshold of four frequencies (0.5, 1, 2, 4 kHz); CI, cochlear implant (CI512, Cochlear, Sydney, Australia); CPA, cerebellopontine angle; IAC, internal auditory canal; m/f, male/female; R/L, right/left; WRS_{max}, maximum number of monosyllabic words understood (in %); WRS_{65}, percentage of words understood (numbers and monosyllables separately tested).

Patients 1 to 4 have been reported in a previous case series (16) and case report (19), as referenced in the ''Procedure'' column of this table.

of the resection of intralabyrinthine schwannomas with intracochlear location or extension, and the short-term functional outcome after cochlea implantation.

MATERIALS AND METHODS

Between 2011 and 2017, 10 patients underwent surgery for removal of an ILS with intracochlear location in our university department (tertiary referral center) (Table 1). Eight patients received a cochlear implant (Model Nucleus CI512, Cochlear, Sydney, Australia) in a single stage procedure together with tumor resection. Two patients opted for insertion of a dummy electrode carrier (insertion test device, MedEl, Innsbruck, Austria), understanding and explicitly asking for it, although this is an off-label procedure. In one case (patient 2) the patient did not want a cochlear implant for personal reasons but wanted to keep the option of a possible cochlear implantation in the

future. The other patient (patient 3) wanted to keep optimal conditions for repeated MRI scans for follow up for residual or recurrent tumor before deciding about a cochlear implantation in the future. Surgery was performed in all patients under general anesthesia.

RESULTS

Figures 1 and 2 show the surgical removal of an intracochlear ILS via subtotal cochleoectomy and cochlear implantation. After opening the cochlear capsule, the tumor was identified and the cochlear turns were subsequently opened following the tumor until the scalar lumina were free of tumor. A cochlear implant electrode carrier (Model Nucleus CI512, Cochlear, Sydney, Australia) was placed around the preserved basal part of the modiolus. The bony arch (*) of the round window and pieces of cartilage were used to stabilize the position of the cochlear implant electrode carrier. In cases with a subtotal cochleoectomy, the defect was closed with a cartilage-perichondrium compound transplant with cartilage island and bone pâté (Figs. 1E–G and 2F). In two patients, an additional labyrinthectomy was necessary due to additional tumor location in the vestibule (patients 6 and 9 in Table 1). In one case (patient 9 in Table 1), a monofilic suture was placed through the tumor via an opening in the basal and in the second turn. After placing two knots at the end of the suture, the tumor was removed by pulling the monofilic strand backwards (Fig. 2G and H).

The surgical approach was through a retroauricular incision and involved a simple mastoidectomy, and, in cases with cochlear implantation, a posterior tympanotomy. Cochlear tumor resection was done through the external ear canal. Apart from the patients were the tumor could be removed through an extended round window approach or a partial cochleoectomy of the anterior parts of the basal and middle turn (patients 1 and 2 in Table 1), the incus had to be removed to get access to the cochlea. The chorda tympani could be preserved only in some cases. The vulnerable structures in the vicinity of the surgical field that need to be considered are the facial nerve (Figs. 1B and 2B, D, E), the internal carotid artery (Fig. 2B), and the modiolus (Figs. 1C and 2D, E). There were no cases of postoperative infection or facial paralysis. Patients without preoperative complete loss of peripheral vestibular function in the tumor ear (i.e., all patients apart from patients 6 and 9 in Table 1) suffered from postoperative vertigo with a duration from some days to some weeks. No patients suffered from vertigo from the 6th week follow up examination onward. One patient needed revision surgery due to a fistula from the ''neochochlea'' and due to a small tympanic membrane perforation. This patient, however, was extremely motivated and started exercising already on the 2nd day after the first surgery, repeatedly climbing stairs to the 7th floor of the hospital, which might have contributed to the fistula. The fistula was successfully closed through an endaural approach with tragal perichondrium and TachoSil sealant matrix (Takeda Austria-GmbH, Linz, Austria). The cochlear implant remained ''untouched'' without any electrophysiological or functional changes. The small tympanic membrane defect was closed in the same session with tragal perichondrium. In all but one case, surprisingly good audiological performances with the cochlear implant were observed already after a 3 months follow up period (Table 1).

DISCUSSION

Like ''classical'' vestibular schwannomas (acoustic neuromas) located in the internal auditory canal and the cerebellopontine angle, intralabyrinthe schwannomas can be managed with a ''wait-and-test-and-scan'' strategy, with radiotherapy or with surgical tumor resection with or without cochlear implantation (3,13–19). Alternative management strategies were, therefore, extensively and repeatedly discussed with the patients before surgery and within the interdisciplinary hearing implant board of our hearing implant center. Patients were offered surgery only if there was a strong wish for surgical tumor removal and/or hearing rehabilitation with a cochlear implant.

The extent of the surgical resection, i.e., extended round window approach, partial or subtotal cochleoectomy with or without labyrinthectomy, was based on tumor location and extension. Similar ''circum-modiolar drill-out'' procedures have been applied in cases of labyrinthitis ossificans (20–23). In patients with intralabyrinthine schwannomas, however, there is no ossification of the fluid spaces of the cochlea, and the tumor could be resected from the bony labyrinth after removal of parts of the cochlear capsule. Therefore, the term ''cochlear drill-out'' was not chosen here.

Although recovery from moderate to mild hearing loss after surgery has been described for an intralabyrinthine schwannoma with vestibular location (24), hearing preservation surgery for tumors with intracochlear location, even if small, is not possible due to the extent of cochlear trauma (Figs. 1 and 2). Thus, only cochlear implantation can restore hearing on the affected side. A perimodiolar (contour) electrode carrier was chosen to ensure electrode placement close to the remaining spiral ganglion cells in the stump of the modiolus (Figs. 1D and 2E). Leaving a bony arch of the former round window, if possible, helps to keep the electrode carrier in place (Fig. 2E, *). A dummy electrode was used in two patients according to their explicit wish, and patients were informed and consented to the off-label use in these cases. Due to the expected fibrosis and shrinkage of tissue used for ''reconstruction'' around the thin electrode dummy, the success of a later exchange to a cochlear implant electrode carrier, however, is uncertain. Indeed, initial observations with a staged approached for surgical tumor resection and later cochlear implantation, although limited to very few patients, found a poorer prognosis for hearing with CI in these cases. Thus, the reported single stage approach appears to have some advantages with respect to hearing outcome compared with a staged approach (13).

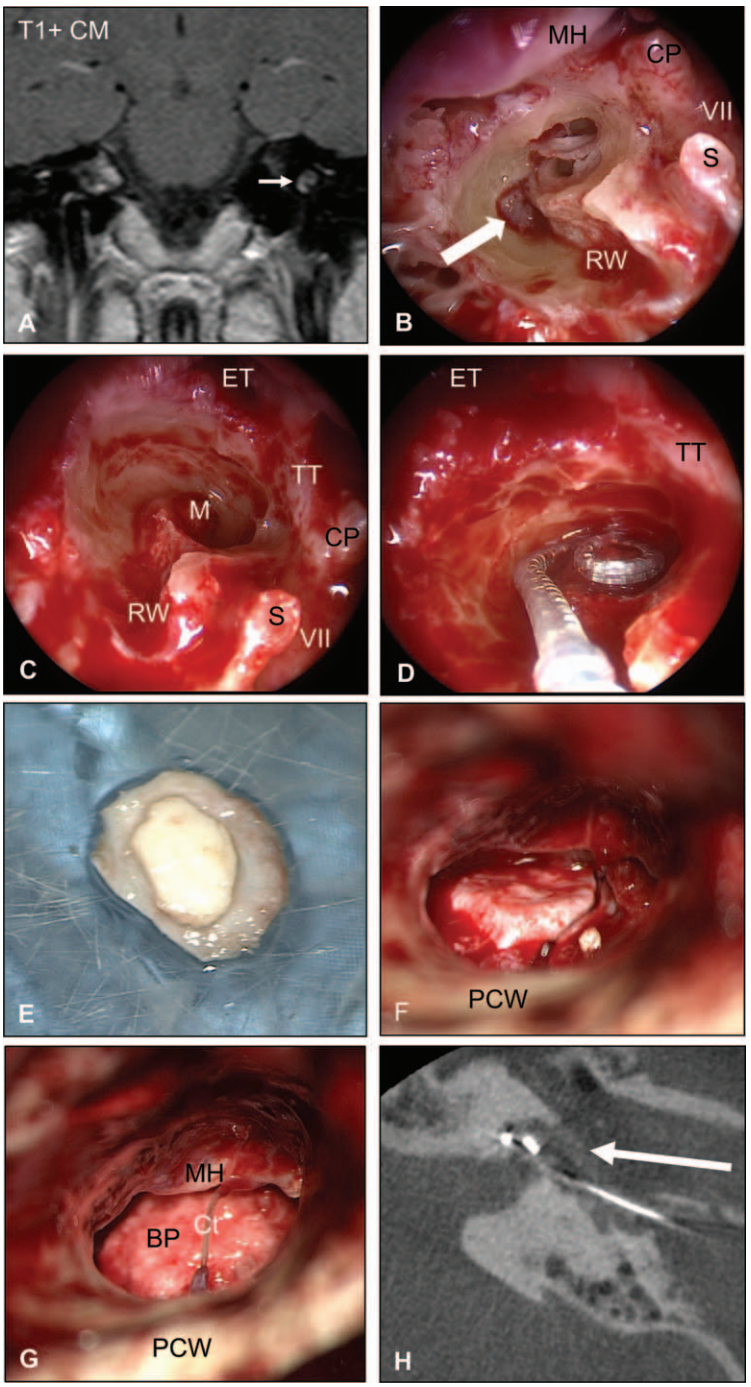

FIG. 1. Surgical removal of an intracochlear ILS via subtotal cochleoectomy and cochlear implantation in a left ear (patient 8 from Table 1): *A*, MRI (coronal, T1-weighted image with contrast medium) showing the tumor (*arrow*) in the basal and partially in the middle turn. *B*, Tumor (*arrow*) in the opened basal turn. The spiral osseous lamina and the organ of Corti are seen in the opened, tumor-free middle and apical turn. *C*, Subtotal cochleoectomy with no signs of remaining tumor. *D*, Cochlear implant electrode carrier placed around the preserved basal part of the modiolus. *E*, Cartilage-perichondrium compound transplant with cartilage island. *F*, Closure of the subtotal cochleoectomy defect with the cartilage-perichondrium compound transplant and bone pâté (*G*). *H*, Postoperative, axial cone beam CT demonstrating the surgical approach to the tumor (*arrow*) and electrode carrier position. BP indicates bone pâté; CP, cochleariform process; Ct, chorda tympani; CT, computed tomography; ET, Eustachian tube orifice; ILS, intralabyrinthine schwannoma; M, modiolus; MH, malleus handle; PCW, posterior canal wall; RW, former round window area; S, stapes head; T1 + CM: T1-weighted image with contrast medium; TT, tensor tympani muscle; VII: facial nerve. *B*, *C*, *D*: endoscopic view (0 degree, 3 mm).

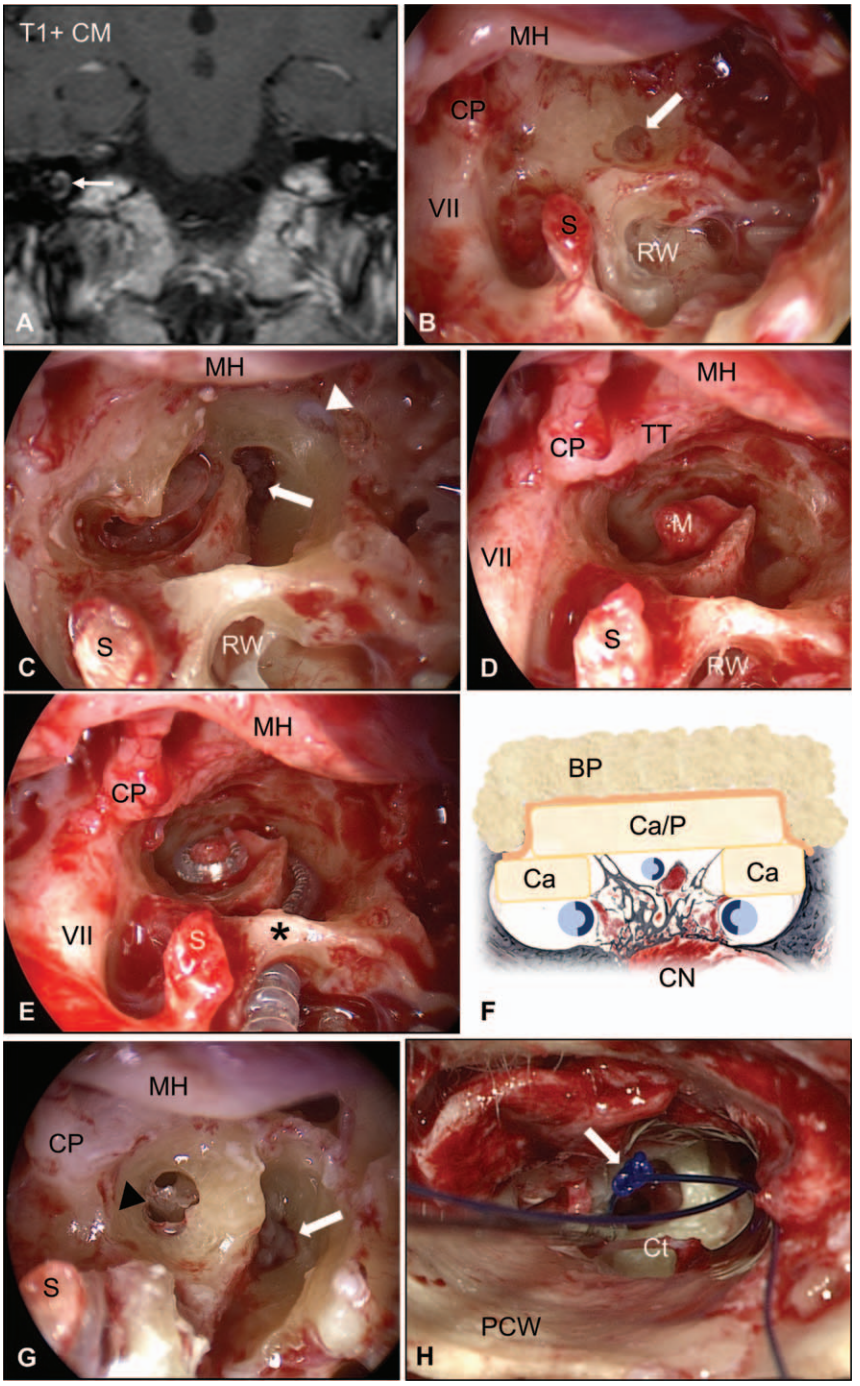

FIG. 2. *A–F*, Surgical removal of an intracochlear ILS via subtotal cochleoectomy and cochlear implantation in a right ear (patient 10 from Table 1): *A*, MRI (coronal, T1-weighted image with contrast medium) showing the tumor (*arrow*) in the basal and partially in the middle turn. *B*, Opening of the cochlear capsule in the anterior part of the basal turn showing the tumor (*arrow*). *C*, The spiral osseous lamina and the organ of Corti are seen in the opened, tumor-free middle turn. *Arrow*: tumor in basal turn. *White arrow head*: internal carotid artery. *D*, Subtotal cochleoectomy with no signs of remaining tumor; *E*, cochlear implant electrode carrier placed around the preserved parts of the modiolus. The bony arch (*) of the round window stabilizes the position of the cochlear implant electrode carrier. *F*, Schematic of the "cochlear reconstruction" through closure of the subtotal cochleoectomy defect with a cartilage-perichondrium compound transplant and bone pâté. The cochlear implant carrier is outlined in light/dark blue. *G*, *H*, Example for the "pull-trough-technique" for removal of an intracochlear tumor (patient 9 from Table 1). A monofil suture is placed through the tumor via an opening in the basal and in the middle turn (*black arrow head*). After placing two knots at the end of the suture, the tumor can be removed by pulling the monofil strand backwards. In these cases, control for complete intrascalar tumor removal appears less reliable then after subtotal cochleoectomy. BP indicates bone pâté; Ca, cartilage (Ca/P with perichondrium); CN, cochlear nerve; CP, cochleariform process; Ct, chorda tympani; ILS, intralabyrinthine schwannoma; M, modiolus; MH, malleus handle; PCW, posterior canal wall; RW, round window; S, stapes head; T1 + CM, T1-weighted image with contrast medium; TT, tensor tympani muscle; VII, facial nerve; *B*, *C*, *D*, *E*, *G*: endoscopic view (0 degree, 3 mm).

An alternative method to ''subtotal cochleoectomy'' for tumor removal, a ''pull-through-technique'', has been applied in patient 9 of this case series (Fig. 2G and H). Tumor resection through ''an expanded cochleostomy of the basal turn and an additional cochleostomy of the second turn'' was previously described by Aschendorff et al. (13). Control for complete intrascalar tumor removal, however, especially in the scalar segments medially to the modiolus, appears less reliable then after subtotal cochleoectomy. We considered this small risk acceptable, since in this patient with an initially likely transmodiolar growth (patient 9 in Table 1), some minimal tumor remnants also most likely remained within the modiolus.

Insertion of a cochlear implant without tumor removal, i.e., insertion through the tumor mass in the cochlea, has also been suggested as a way of successful hearing rehabilitation in selected cases (14).

To our knowledge, there is no reported experience with radiation therapy of intracochlear schwannomas. Hearing rehabilitation with cochlear implants after radiotherapy of ILS, however, appears very unlikely due to the expected damage to the neural structures (cochlear spiral ganglion cells). The disadvantage of a ''wait-and-test-and-scan'' strategy includes transmodiolar tumor progress into the internal auditory canal (IAC). A transotic-translabyrinthine tumor removal in these cases would not allow rehabilitation with a CI (13,14,16).

Due to the accessibility of intralabyrinthine schwannomas through the external ear canal and the mastoid, which are standard access pathways for the otosurgeon, in view of the good audiological short-term hearing results after CI, and the limited impact on vestibular function, surgical removal of intracochlear ILS is considered a promising therapeutic strategy. It will be of importance, however, to observe the long-term outcome in these patients, and if for instance fibrosis and ossification will become a problem following this approach. Initial results after 2-year follow up in patient 1 (with an extended round window approach) and results after 6 months in patients with subtotal or partial cochleoectomy (Table 1) are promising so far. If hearing rehabilitation in these patients is successful also in the long term, this would add interesting aspects to the discussion about the role of ''soft surgery approaches'' and other technical issues in cochlear implantation.

Future challenges include MRI-follow-up for tumor recurrence or residual tumor growth. Although a complete removal cannot be guaranteed by the surgical techniques above, and microscopic tumor cells especially in the modiolus may remain, this may not lead to situations requiring treatment in the future, based on the very slow growth tendency of ILS (25). In addition, initial observations have already demonstrated that imaging of the inner ear, the internal auditory canal, and the cerebellopontine angle is possible even after cochlear implantation (14,26).

REFERENCES

1. Dubernard X, Somers T, Veros K, et al. Clinical presentation of intralabyrinthine schwannomas: a multicenter study of 110 cases. *Otol Neurotol* 2014;35:1641–9.
2. Frisch CD, Eckel LJ, Lane JI, et al. Intralabyrinthine schwannomas. *Otolaryngol Clin North Am* 2015;48:423–41.
3. Gosselin E, Maniakas A, Saliba I. Meta-analysis on the clinical outcomes in patients with intralabyrinthine schwannomas: conservative management vs. microsurgery. *Eur Arch Otorhinolaryngol* 2016;273:1357–67.
4. Jerin C, Krause E, Ertl-Wagner B, et al. Clinical features of delayed endolymphatic hydrops and intralabyrinthine schwannoma: An imaging-confirmed comparative case series. *HNO* 2017;65:41–5.
5. Slattery EL, Babu SC, Chole RA, et al. Intralabyrinthine schwannomas mimic cochleovestibular disease: symptoms from tumor mass effect in the labyrinth. *Otol Neurotol* 2015;36:167–71.
6. Van Abel KM, Carlson ML, Link MJ, et al. Primary inner ear schwannomas: a case series and systematic review of the literature. *Laryngoscope* 2013;123:1957–66.
7. Arndt S, Aschendorff A, Laszig R, et al. Comparison of pseudobinaural hearing to real binaural hearing rehabilitation after cochlear implantation in patients with unilateral deafness and tinnitus. *Otol Neurotol* 2011;32:39–47.
8. Arts RA, George EL, Stokroos RJ, et al. Review: cochlear implants as a treatment of tinnitus in single-sided deafness. *Curr Opin Otolaryngol Head Neck Surg* 2012;20:398–403.
9. Rahne T, Plontke SK. Functional result after cochlear implantation in children and adults with single-sided deafness. *Otol Neurotol* 2016;37:e332–40.
10. van Zon A, Peters JP, Stegeman I, et al. Cochlear implantation for patients with single-sided deafness or asymmetrical hearing loss: a systematic review of the evidence. *Otol Neurotol* 2015;36:209–19.
11. Vermeire K, Van de Heyning P. Binaural hearing after cochlear implantation in subjects with unilateral sensorineural deafness and tinnitus. *Audiol Neurootol* 2009;14:163–71.
12. Arndt S, Laszig R, Aschendorff A, et al. Cochlear implant treatment of patients with single-sided deafness or asymmetric hearing loss. *HNO* 2017;65 (Suppl 2):98–108.
13. Aschendorff A, Arndt S, Laszig R, et al. Treatment and auditory rehabilitation of intralabyrinthine schwannoma by means of cochlear implants: English version. *HNO* 2017;65 (Suppl 1):46–51.
14. Carlson ML, Neff BA, Sladen DP, et al. Cochlear implantation in patients with intracochlear and intralabyrinthine schwannomas. *Otol Neurotol* 2016;37:647–53.
15. Kronenberg J, Horowitz Z, Hildesheimer M. Intracochlear schwannoma and cochlear implantation. *Ann Otol Rhinol Laryngol* 1999;108:659–60.
16. Plontke SK, Rahne T, Pfister M, et al. Intralabyrinthine schwannomas: surgical management and hearing rehabilitation with cochlear implants. *HNO* 2017;65 (Suppl 2):136–48.
17. Schutt CA, Kveton JF. Cochlear implantation after resection of an intralabyrinthine schwannoma. *Am J Otolaryngol* 2014;35:257–60.
18. Shupak A, Holdstein Y, Kaminer M, et al. Primary solitary intralabyrinthine schwannoma: A report of 7 cases and a review of the literature. *Ear Nose Throat J* 2016;95:481–91.
19. Plontke SK, Kosling S, Pazaitis N, et al. Intracochlear schwannoma: tumor removal via subtotal cochleoectomy and partial cochlear reconstruction with preservation of semicircular canal function. *HNO* 2017;65 (Suppl 2):158–62.
20. Balkany T, Bird PA, Hodges AV, et al. Surgical technique for implantation of the totally ossified cochlea. *Laryngoscope* 1998;108:988–92.
21. Balkany T, Gantz B, Nadol JB Jr. Multichannel cochlear implants in partially ossified cochleas. *Ann Otol Rhinol Laryngol Suppl* 1988;135:3–7.
22. Rauch SD, Herrmann BS, Davis LA, et al. Nucleus 22 cochlear implantation results in postmeningitic deafness. *Laryngoscope* 1997;107:1606–9.

23. Wanna GB, Carlson ML, Blachon GS, et al. Implantation of the completely ossified cochlea: an image-guided approach. *Otol Neurotol* 2013;34:522–5.
24. Lee JJ, Lim HJ, Park HY, et al. Recovery of hearing after surgical removal of intralabyrinthine schwannoma. *Laryngoscope* 2015;125:1968–71.
25. Tieleman A, Casselman JW, Somers T, et al. Imaging of intralabyrinthine schwannomas: a retrospective study of 52 cases with emphasis on lesion growth. AJNR. *Am J Neuroradiol* 2008;29:898–905.
26. Todt I, Rademacher G, Mittmann P, et al. Postoperative imaging of the internal auditory canal: visualization of active auditory implants. *HNO* 2017;65 (Suppl 2):81–6.