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Revised Classification of Inner Ear Schwannomas

*Stefan K. Plontke, †‡§Simon K.W. Lloyd, ‡§Simon R.M. Freeman, ||Sabrina Kösling, ¶Christoph Arnoldner, **Nigel Biggs, ††Daniele Borsetto, ‡‡Samuel Gubbels, §§Janette Hess-Erga, ||||Ja-Won Koo, ¶¶Christine M. Lohse, ***John P. Marinelli, †††Riccardo di Micco, ‡‡‡Ashley M. Nassiri, *Torsten Rahne, *Jonas Scheffler, §§§,||||||Per Cayé-Thomasen, and ***Matthew L. Carlson

**Department of Otorhinolaryngology, Head & Neck Surgery, University Medicine Halle, Halle (Saale), Germany; †Faculty of Biology, Medicine and Health, Division of Cancer Sciences, University of Manchester, Manchester M13 9PL, UK; ‡Department of Otolaryngology, Manchester Royal Infirmary, Manchester University Hospitals NHS Foundation Trust, Manchester Academic Health Science Centre, Manchester M13 9WL, UK; §Department of Otolaryngology Salford Royal Hospital, Northern Care Alliance, Manchester M6 8HD, UK; ||Department of Radiology, University Medicine Halle, Halle (Saale), Germany; ¶Christian Doppler Lab for Inner Ear Research, Department of Otorhinolaryngology, Head and Neck Surgery, Medical University of Vienna, Vienna, Austria; **Department of Otolaryngology, Head & Neck Surgery, St Vincent's Hospital, Sydney, Australia; ††Department of ENT Surgery, Addenbrookes Hospital, Cambridge University Hospitals NHS Foundation Trust, Cambridge, UK; ‡‡Departments of Otolaryngology and Neurosurgery; University of Colorado, School of Medicine; Aurora, Colorado, USA; §§Department of otorhinolaryngology, Head and Neck Department, Haukeland University Hospital, Bergen, Norway; ||||Ja-Won Koo; Department of Otorhinolaryngology, Seoul National University Bundang Hospital, Seoul National University College of Medicine, Seoul, South Korea; ¶¶Department of Quantitative Health Sciences, Mayo Clinic, Rochester, Minnesota, USA; ***Department of Otolaryngology–Head and Neck Surgery, Mayo Clinic, Rochester, Minnesota, USA; †††Hannover Medical School, ENT Clinic, Hannover, Germany; ‡‡‡University of Colorado Anschutz School of Medicine, Aurora, Colorado, USA; §§§Copenhagen Hearing and Balance Center, Department of ORL HNS and Audiology, University Hospital Rigshospitalet, Copenhagen, Denmark; and ||||||Faculty of Health Sciences, Copenhagen University, Copenhagen, Denmark*

ABSTRACT: Over the past two decades, there has been increasing interest in the diagnosis and management of schwannomas of the inner ear including hearing rehabilitation with cochlear implants. However, tumor nomenclature and classification within the literature have been variable and oftentimes cumbersome. The term "intralabyrinthine schwannoma" is in common use when describing these tumors but is a potential source of confusion given that people often use the term "labyrinth" or "labyrinthine" to refer to the vestibular component of the inner ear only (i.e., labyrinthectomy or the translabyrinthine approach). During the Ninth Quadrennial Conference on Vestibular Schwannoma and Other Cerebellopontine Angle Lesions in Bergen, Norway, in May 2023, a multidisciplinary group of conference participants met and discussed issues pertaining to current terminology and classifications to enhance clarity and to reflect recent advances in tumor management and hearing rehabilitation.

Although a variety of terms have been previously used to describe inner ear schwannomas, consensus was achieved on the term "inner ear schwannoma (IES)" to describe eighth nerve schwannomas of the cochlea, vestibule, or semicircular canals. Subgroups under this term comprise intravestibular, intracochlear, or intravestibulocochlear inner ear schwannomas (low complexity tumors), inner ear schwannomas with transfundal extension into the internal auditory canal but without modiolar involvement (intermediate complexity tumors), and inner ear schwannomas with transfundal extension with modiolar involvement (high complexity tumors).

The details of the recommendations for an updated and simplified tumor nomenclature centered around tumor control and hearing rehabilitation with cochlear implantation are presented.

Key Words: Acoustic neuroma—Cochlear implant—Cochlear implantation—Inner ear schwannoma—Intracochlear schwannoma—Intralabyrinthine schwannoma—Neurotology—Schwannoma—Vestibular schwannoma.

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

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INTRODUCTION

During the past two decades, there has been increasing interest in the diagnosis and management of schwannomas of the inner ear (1). Because of better imaging technology, an increased awareness of the entity itself, and a high desire to find causes for inner ear diseases and symptoms like sudden hearing loss and vertigo, the incidence of this tumor seems to have increased (2).

In addition, because cochlear implantation (CI) offers the opportunity for hearing rehabilitation in many cases of schwannomas of the inner ear (3–8) and CI is an accepted and often reimbursed rehabilitation for single-sided deafness in many countries, the topic has become increasingly clinically practical.

The term "intralabyrinthine schwannoma," is in common use when describing these tumors but is a potential source of confusion given that people often use the term "labyrinth" or "labyrinthine" to refer to the vestibular component of the inner ear (i.e., the vestibular labyrinth) only. This confusion may arise in part from use of standard otologic terms, such as labyrinthectomy or the translabyrinthine approach, which only refer to the removal of the vestibular portion of the inner ear. Use of a more generic term, such as "inner ear schwannoma," (IES) would raise awareness that these tumors can occur in all areas of the inner ear, not just the vestibular portion. Furthermore, these tumors can grow into more than one area of the inner ear and can extend beyond the structures of the labyrinth. Finally, several terms used in current classifications are cumbersome and nonspecific, such as transotic and translabyrinthine.

Therefore, during the Ninth Quadrennial Conference on Vestibular Schwannoma and Other Cerebellopontine Angle Lesions in Bergen, Norway, in May 2023, a multidisciplinary group of conference participants met and discussed issues pertaining to current terminology and classifications. The participants agreed on the necessity for revised nomenclature and classification to enhance clarity and to reflect recent advances in tumor management and hearing rehabilitation. Nomenclature and classification should be clear, simple, practical, memorable, and management oriented. It should also be easy to use to facilitate interdisciplinary communication (e.g., with radiologists and neurosurgeons). The recommendations for terminology and classification produced at this meeting are presented herein.

EVOLUTION OF TERMINOLOGY AND CLASSIFICATION

The first descriptions of IES are found in case reports as early as 1917. Mayer described "a case of multiple tumors in the distal branches of the acoustic nerve," and in the same issue of the same journal, Nager described a "neurofibroma of the cochlear spiral." In both cases, the tumors were found in the cochlear scalae and in the modiolus (9,10). Jorgensen in 1962 (11) published histological images of an "intracochlear neurinoma" in the scala tympani of the basal turn, which occupied "the entire lumen," penetrated "the wall toward the modiolus," and filled "the greater part of Rosenthal's canal but without involvement of the internal auditory meatus." In 1999, Kronenberg et al. (12) described a successful (staged) CI after discovering an "intracochlear schwannoma" in the basal turn of the cochlea during surgery.

In 2004, Kennedy et al. (13) proposed an initial classification of "intralabyrinthine schwannomas" that included intravestibular (vestibule ± semicircular canals), intracochlear (cochlea), intravestibulocochlear (vestibule and cochlea), transmodiolar (cochlea and internal auditory canal [IAC]), transmacular (vestibule and IAC), transotic (middle ear and vestibule/cochlea and IAC), and tympanolabyrinthine (middle ear and vestibule/cochlea) locations and extensions. Tieleman et al. (14) additionally specified tumor locations in the cochlea (basal, second, or apical parts or combinations, and scala tympani, scala vestibuli, or both) and in the vestibular labyrinth (anterior/posterior vestibule with or without involvement of semicircular canals [SCC]) without proposing this as a separate classification.

In 2011, Massager et al. (15) proposed an anatomically based classification (I–IV with subclasses a–c) based on seven cases, but with little clinical or surgical reference. In an extended case series, Salzmann et al. (13,16) revised the initially proposed classification from their group by excluding the "tympanolabyrinthine class [middle ear and vestibule/cochlea] … due to the fact that there were no observed cases fitting the description and there was redundancy with the transotic subtype." Van Abel et al. (17) included the "tympanolabyrinthine" class again, introduced a "translabyrinthine" class (vestibulocochlear and IAC), and differentiated between "transotic" and "transotic with cerebellopontine angle (CPA) involvement". In contrast, in a large multicenter analysis, Dubernard et al. (18) arbitrarily excluded lesions with relevant IAC extension and all IES with extension into the CPA.

PROPOSED CLASSIFICATION OF INNER EAR SCHWANNOMAS

The working group agreed that the core term "inner ear schwannoma" (IES) provides an accurate and clearer description of the pathology. Broadly, these tumors are managed with wait-and-scan, microsurgical resection, or less commonly, stereotactic radiosurgery. We discussed the potential for confusion between IES and vestibular schwannoma (VS). The term inner ear schwannoma immediately highlights involvement of this structure (the inner ear), and the addition of the term transfundal further serves this purpose when there is tumor within the IAC and CPA, which could potentially be confused with VS radiologically. In addition, CI can be performed with or without "treatment" of the tumor in many cases. The group believes that tumors should be divided according to the complexity of excision taking into consideration the potential for hearing rehabilitation with CI.

For tumors extending from the inner ear into the IAC and even into the CPA, the group recommends using the terms "transfundal IES without modiolar involvement" or "transfundal IES with modiolar involvement" depending on the route of extension from the inner ear to the fundus of the IAC. Based on the aim of creating a clear, simple,

practical, memorable, and management-oriented classification that allows for easy interdisciplinary communication (e.g., with radiologists and neurosurgeons), the group believes that terms like "transmacular" that are uncommonly used in clinical care should be avoided, whereas the fundus of the IAC is a well-known anatomical landmark both radiologically and surgically. Furthermore, the involvement of the modiolus of the cochlea is the main factor in the complexity of the treatment of these tumors, especially regarding hearing rehabilitation with CI, an aspect that is highlighted with the proposed classification.

Lower Complexity

Inner ear schwannomas confined to the inner ear can either develop within the cochlea or in the vestibule with or without partial or complete involvement of the SCC. The group determined that the terms "intravestibular IES" and "intracochlear IES" should be used for tumors confined to the vestibular portion of the labyrinth or the cochlea, respectively (Fig. 1, A and B). The group discussed adding the prefix "canalo-" for tumors also involving the SCC. Because an extension from the vestibule to the SCC seems of no relevance for management and the term "intracanalovestibular" is quite cumbersome, the group suggested that it was not necessary to have a specific term for this type of tumor. If the tumor involves both the cochlea and the vestibular labyrinth, it would be termed "intravestibulocochlear IES" (Fig. 1C).

Within this subset of IES, tumors isolated to the vestibule, with or without involvement of the SCC, represent the most straightforward group as complete resection can be achieved via transmastoid labyrinthectomy. Surgical excision of intracochlear IES would involve complete cochlectomy (if CI is not being considered) or, depending on the extent of disease, an extended cochleostomy or partial or subtotal cochlectomy with preservation of the modiolus for hearing rehabilitation with CI and the possibility of preservation of vestibular function in most cases (19,20). In these cases, a transcanal approach to the cochlea can be used or, in some cases, a transmastoid with facial recess approach, or blind-sac closure of the ear canal with or without subtotal petrosectomy, depending on the location and extent of the tumor. For "intravestibulocochlear IES," labyrinthectomy combined with one of the above cochlear approaches is indicated.

Intermediate Complexity

These tumors have extension from the vestibule into the IAC and possibly the CPA. They might involve the cochlea, but the modiolus and spiral ganglion cells are not involved and CI with complete tumor removal is thus possible. However, surgical resection is more challenging than in lower complexity tumors because the subarachnoid barrier is entered, and the cochlear nerve can be in contact with the IAC portion of the tumor. These tumors would be described as "transfundal IES without modiolar involvement" (formerly referred to as "transmacular") (Figs. 1D and 2A). These tumors can be surgically managed through a translabyrinthine approach with or without the cochlear approaches outlined above, depending on tumor extent in the cochlea.

High Complexity

These tumors have extension from the cochlea into the IAC through the modiolus (Figs. 1E and 2B), with or without involvement of the vestibule. These tumors should be described by the term "transfundal IES with modiolar involvement" (formerly referred to as "transmodiolar"). Hearing rehabilitation with CI is significantly more

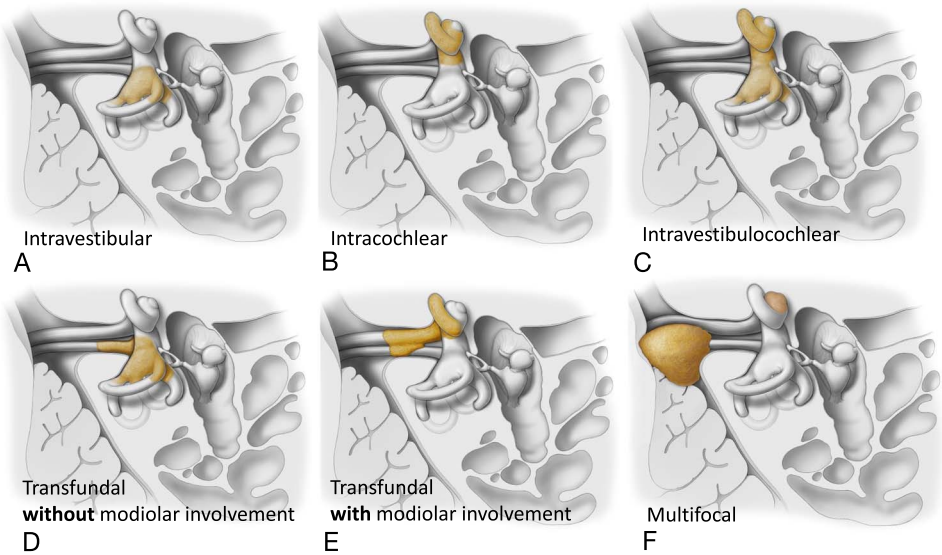

FIG. 1. *A–F*, Schematic drawing of various locations for inner ear schwannomas (IES): (*A*) intravestibular IES, (*B*) intracochlear IES, (*C*) intravestibulocochlear IES, (*D*) transfundal IES without modiolar involvement (formerly named as transmacular IES), (*E*) transfundal IES with modiolar involvement (formerly named as transmodiolar IES), (*F*) multifocal schwannoma (intracochlear IES and cerebellopontine angle schwannoma). Drawings by Hans Jörg Schütze, Köln, Germany in collaboration with the first author.

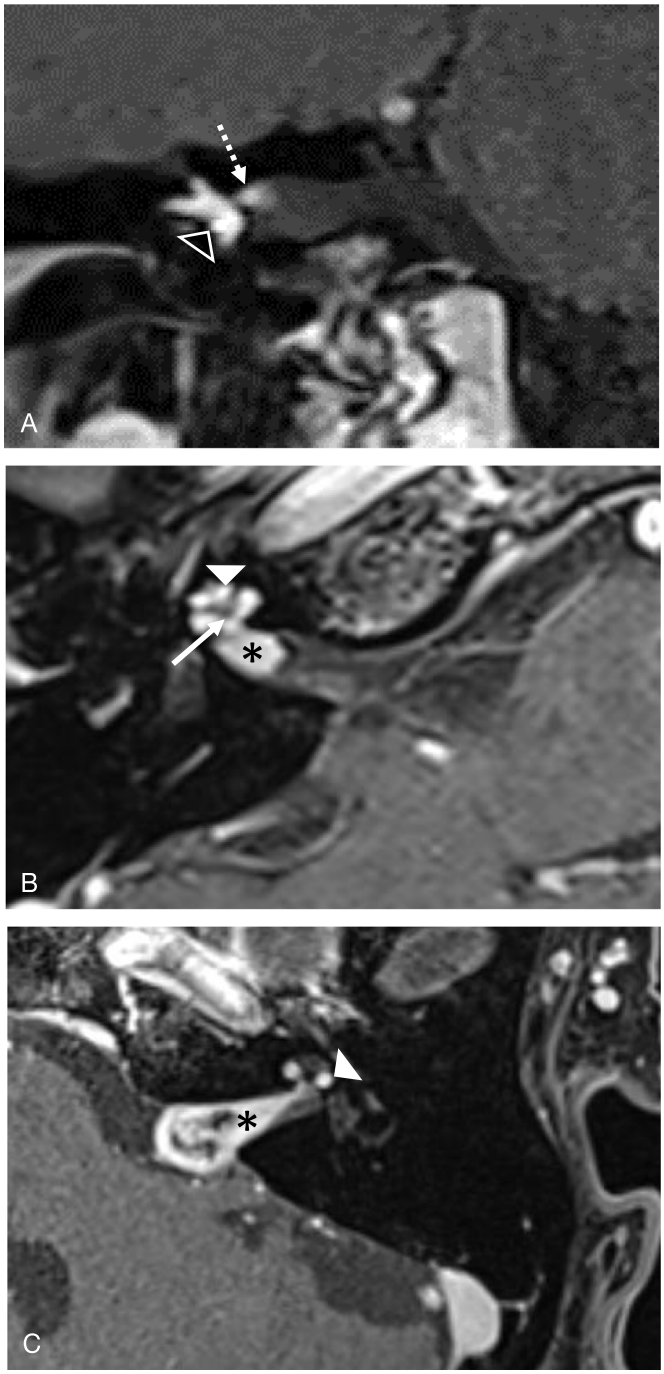

FIG. 2. *A–C*, Transfundal inner ear schwannomas (IES). *A*, Transfundal IES without modiolar involvement: MRI (coronal, T1-w + Gd) showing tumor in the right vestibule (*arrowhead*), in the ampullary ends of the superior and lateral semicircular canals and extending along the superior vestibular nerve into the IAC (dotted arrow). From Plontke et al. (HNO, Springer) (21). *B*, Transfundal IES with modiolar involvement: MRI (right ear, axial, T1-w + Gd) showing tumor in the cochlea (*arrowhead*) with extension through the modiolus (➔) into the IAC (*). *C*, Transfundal IES with extension from the cochlea into the IAC and CPA: MRI (left ear, axial, T1-w + Gd) showing a tumor in the middle turn of the cochlea (*arrowhead*) extending into the IAC (*) and the CPA. Courtesy of Professor Dr. med. Philippe L. Pereira, Center of Radiology, Minimally Invasive Therapies and Nuclear Medicine, Heilbronn, with permission. *B* and *C*, If the tumor involves the inner ear and the internal auditory canal at first presentation, it is usually impossible to determine with any certainty whether the inner ear portion was "primary" or "secondary." IAC indicates internal auditory canal; CPA, cerebellopontine angle; Gd, gadolinium; -w, weighted.

complex if microsurgical tumor resection is pursued. Because the spiral ganglion cells in the modiolus are needed for stimulation with a cochlear implant, complete surgical tumor removal is not possible if the tumor traverses the modiolus. If CI is aimed for, electrode insertion without tumor removal (4,22) or after partial tumor removal (21),

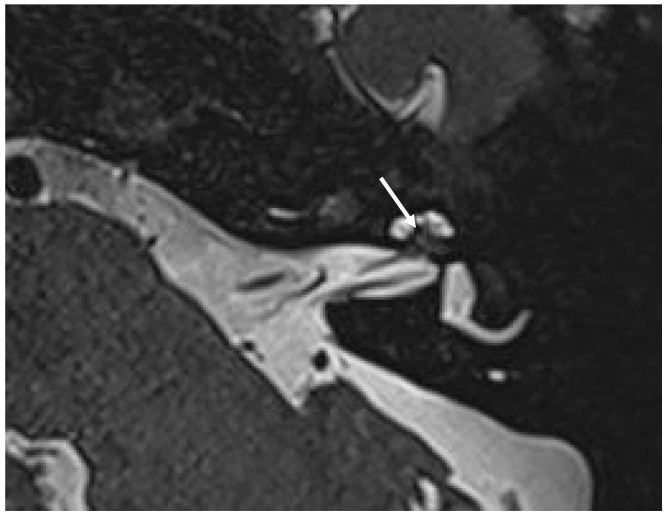

FIG. 3. MRI (left ear, axial, T2-w) showing a small inner ear schwannoma of the middle cochlear turn with modiolar involvement (*arrow*), but without relevant extension into the fundus of the internal auditory canal. -w indicates weighted.

possibly with previous radiotherapy, can be considered. Intracochlear tumors with involvement of the modiolus that have not (yet) extended to the IAC (Figs. 3 and 5B) would also meet these criteria given that complete tumor removal will hinder CI for the above reasons. However, compared with tumors extending further into the IAC or even into the CPA, tumor removal bears no risk to the facial nerve in the IAC.

Involvement of the Cerebellopontine Angle

Further extension of transfundal tumors into the CPA renders more risk to the cochlear nerve, facial nerve, and other surrounding neurovascular structures (Fig. 2C). Tumor size or involvement of the CPA and its extent, respectively, will thus influence and further complicate but not generally change the surgical approach for transfundal tumors. Therefore, a separate category for CPA extension was not felt to be required, and the group recommends simply adding the term "+ CPA extension" to these tumors (e.g., "transfundal IES without or with modiolar involvement + CPA extension") (Fig. 2C).

Involvement of the Middle Ear

Tumor extension from the inner ear to the middle ear (e.g., through the round window membrane, the stapes footplate, or the SCCs, (23)) had been termed "tympanolabyrinthine" in earlier classifications (13,17). However, as this does not describe the extent of the tumor in the inner ear, and middle ear extension is rare and generally has minimal impact on management or outcome, the group recommended describing this additional extension separately (e.g., "intracochlear IES + middle ear extension").

For the same reasons, the terms "transotic" (middle ear + inner ear + IAC) or "transotic + CPA" (middle ear + inner ear + IAC + CPA, (13,16,17)) were considered unnecessary. Their management complexity does not differ from larger transfundal IES.

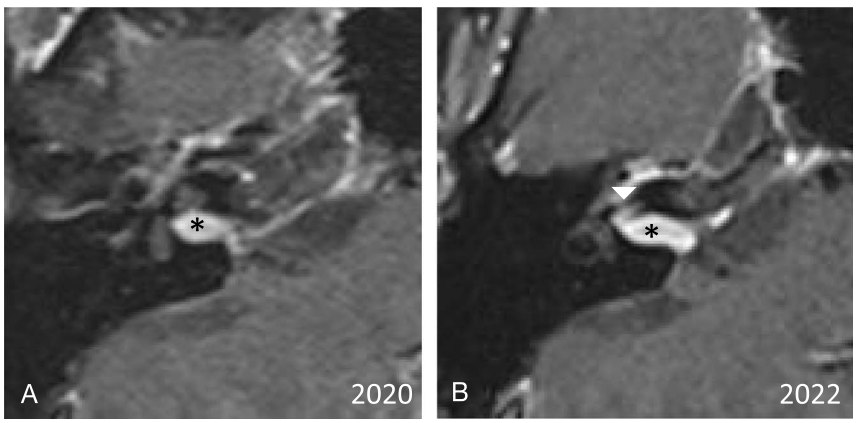

FIG. 4. MRIs (right ear, axial, T1-w + Gd, FS) showing a tumor in the internal auditory canal (*A*) (*) with secondary growth through the modiolus into the middle turn of the cochlea (*arrowhead* in *B*), which is rare in patients without NF2. Gd indicates gadolinium; FS, fat saturation; -w, weighted.

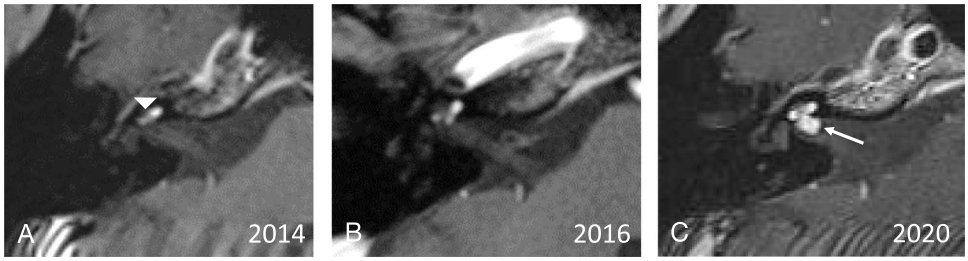

FIG. 5. MRI (right ear, axial, T1-w + Gd) showing an inner ear schwannoma primarily located in the cochlea (*arrowhead*) with secondary growth through the modiolus into the internal auditory canal (*arrow*) during an interval of 6 years. *A* and *B*, Courtesy of Prof. Dr. Ulrike Ernemann, Diagnostic and Interventional Radiology, University Hospital Tübingen, with permission. Gd indicates gadolinium; -w, weighted.

Other Aspects

The term "translabyrinthine" was not part of the initial Kennedy classification but later introduced by Van Abel et al. (13,17). It described intravestibulocochlear IES with involvement of the entire fundus of the IAC. Given that it may be difficult to determine how growth occurred from the inner ear to the IAC or vice versa (i.e., through the modiolus, the macula cribrosa, or both in some cases) and a "translabyrinthine" tumor cannot be managed with a "translabyrinthine" surgical approach, the group felt that the term "translabyrinthine" should be replaced with the more precise anatomical term "transfundal."

If there are multiple tumors present, for example, sporadic bilateral IES (24) or multifocal inner ear and IAC/CPA schwannomas in non–NF2-related schwannomatosis (NF2) patients (Fig. 1F) (25,26), each should be described individually, and in these patients, as in cases of NF2 (27), the genetic/molecular background should be described if known.

Primary Versus Secondary Inner Ear Schwannomas

The group also discussed the necessity for using the adjectives "primary" and "secondary" IES depending on whether the tumor originates from the inner ear and extends beyond its confines or arises outside the inner ear and extends into it. This differentiation was first suggested by Van Abel et al. (17). For schwannomas that are solely located within the confines of the inner ear, "primary" will not add information. For schwannomas that involve the inner ear and the IAC (+/− CPA) at first presentation, it is usually impossible to determine with any certainty whether they are "primary" or "secondary" (Figs. 2 and 3). Tumors that are initially confined to the IAC but grow into the inner ear should be regarded as vestibular schwannomas with inner ear extension (Fig. 4) (25). They are, however, very rare outside of NF2. Tumors that are confined to the inner ear but subsequently grow into the IAC should be regarded as IES (Fig. 5). Even if primary or secondary status is proven, it is likely unimportant for their management. The group therefore agreed that it is generally unnecessary to use the terms "primary" and "secondary."

The proposed classification was recently successfully applied in an international multi-institutional study of 106 patients with inner ear schwannoma and cochlear implantation (28).

CONCLUSION

Consensus was achieved on the term "inner ear schwannoma (IES)" to describe eighth nerve schwannomas of the cochlea, vestibule, or semicircular canals. Subgroups under this term comprise intravestibular, intracochlear, or intravestibulocochlear

TABLE 1. *Revised classification of inner ear schwannomas*

	Tumor Present in[a]			
Classification of IES	Cochlea	Vestibule	SCC	IAC
Intravestibular IES		X	(X)	
Intracochlear IES	X			
Intravestibulocochlear IES	X	X	(X)	
Transfundal IES without modiolar involvement[b]	(X)	X	(X)	X
Transfundal IES with modiolar involvement[c]	X	(X)	(X)	X
+ Involvement of the CPA	+ CPA			
+ Involvement of the middle ear	+ Middle ear			
Bilateral[d]	State bilateral			
Multifocal[d]	Describe locations			

[a] "X" indicates anatomical compartment(s) that must be involved; "(X)" indicates anatomical compartment(s) that may or may not be involved.
[b] Transfundal tumors without modiolar involvement are likely to arise from the vestibule with extension through the macula cribrosa into the IAC. They might have cochlear involvement.
[c] Transfundal tumors with modiolar involvement are likely to arise from the cochlea with extension through the modiolus into the IAC. Additional extension from the cochlea to the vestibule is also possible.
[d] In cases of NF2-related schwannomatosis (mosaicism or full), the genetic/molecular background should be described if known.
CPA indicates cerebellopontine angle; IAC, internal auditory canal; IES, inner ear schwannoma; SCC, semicircular canals.

inner ear schwannomas (low complexity tumors), inner ear schwannomas with transfundal extension into the internal auditory canal but without modiolar involvement (intermediate complexity tumors), and inner ear schwannomas with transfundal extension with modiolar involvement (high complexity tumors). The use of the term "modiolar involvement" best indicates whether or not the region of the spiral ganglion cells that receive the electrical stimuli from a cochlear implant is affected, which is crucial for the choice of treatment strategy with regard to hearing rehabilitation with CI. We recommend using the term "inner ear schwannoma" (IES) instead of "intralabyrinthine schwannoma" and the further detailed classification according to Table 1.

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