

Imaging of the Vestibular Schwannoma

Diagnosis, Monitoring, and Treatment Planning



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KEYWORDS

- Neoplasms • Skull base schwannoma • Vestibular computed tomography
- Magnetic resonance imaging • Cerebellopontine angle tumor • Residual tumor • Neurosurgery
- Hearing disorders

KEY POINTS

- Whilst vestibular schwannomas are the commonest tumour of the cerebello-pontine angle cistern and internal auditory meatus, imaging features suggesting an alternative diagnosis should be identified.
- 3D T2w sequences are a cost effective means to detecting vestibular schwannomas, however gadolinium enhanced imaging is required in particular settings and for equivocal findings.
- Understanding the natural history, optimal measurement and definition of tumour growth helps evaluate for the failure of conservative management and requirement for therapeutic intervention.
- MRI helps decide whether surgery or radiotherapy is the appropriate therapeutic intervention, provides important information for surgical planning and monitors tumours following treatment.

Vestibular schwannomas (VSs) account for 6% to 8% of intracranial tumors and comprise the vast majority of cerebellopontine angle (CPA) cistern and internal auditory meatus (IAM) lesions. Imaging plays a key role in the detection, characterization, and management of these tumors.

IMAGING STRATEGIES

MRI is the standard of care for the detection and follow-up of VSs. Heavily T2-weighted (T2w) 3D acquisitions provide high-spatial-resolution isotropic depiction of the CPA and IAM with a cisternographic effect. These may be based on 3D T2w gradient echo (GRE) or 3D fast spin echo (FSE) techniques (**Table 1**). They are used for the initial detection of VSs and are increasingly used

for the monitoring of tumors. The 3D T2w GRE approach uses steady-state free precession sequences which provide high signal to noise. While banding artifact may result from phase shift errors due to magnetic field inhomogeneities, these are mitigated by acquiring at least two off resonance volumes (eg, fast imaging using steady-state acquisition [FIESTA], constructive interference in steady state [CISS]). The 3D T2w FSE sequences are less affected by such susceptibility artifact; however, they have been prone to blurring in view of the long echo train lengths required. These deficiencies have been addressed by contemporary 3D T2w FSE techniques such as volume, isotropic turbo spin echo acquisition (VISTA), CUBE, and sampling, perfection with application optimized contrasts using different flip angle

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Table 1
Imaging sequences

Sequence Type	GE	Siemens	Philips
3D T2w GRE	3D FIESTA-C	3D CISS	3D -SSFP
Optimised 3D FSE (T1w and T2w) ^a	CUBE	SPACE	VISTA

Abbreviations: FIESTA, fast imaging using steady-state acquisition; SPACE, sampling perfection with application optimized contrasts using different flip angle evolution; SSFP, steady state free precession; VISTA, volume isotropic turbo spin echo acquisition.

^a Variations in factors such as the effective echo time and flip angle are used to influence tissue contrast.

evolution (SPACE). These 3D T2w FSE techniques are characterized by significantly shortened echo spacing, and variable flip angles for the refocusing radio-frequency pulses, so suppressing blurring while reducing flow and chemical shift artifacts.

Thin section pre- and post-gadolinium-enhanced T1-weighted imaging is used to characterize lesions and may be used for the posttreatment follow-up of tumors. Fat-saturated T1w imaging may be a useful adjunct in the setting of a postoperative fat graft. While 2- to 3-mm-thin-section spin echo T1w sequences have traditionally been used to image the posterior fossa, 3D T1w sequences are now considered optimal. In particular, 3D T1w FSE sequences benefit from reduced vascular enhancement and susceptibility effects compared with 3D T1w GRE sequences (see **Table 1**).

Innovative MRI techniques such as "Slice Encoding for Metal Artifact Correction (SEMAC)" may also be applied in the setting of patients with MRI conditional auditory implants to improve visualization of the CPA and IAM. CT plays a role in patients with contraindications to MRI and may occasionally be used for presurgical planning.

IMAGING APPEARANCES AND DIFFERENTIAL DIAGNOSIS OF THE CPA CISTERN VESTIBULAR SCHWANNOMA

A CPA cistern VS usually extends from the IAM through the porus acusticus before expanding into the CPA with an "ice cream cone" configuration. MRI reveals a T1w isointense and T2w hyperintense lesion. While smaller schwannomas usually show homogenous enhancement, up to 40% are seen to enhance heterogeneously with 5% to 15% demonstrating cystic change (**Fig. 1**).¹ The presence of tumoral cysts is important to document because they predispose to rapid growth, increase surgical morbidity, and reduce the efficacy of radiotherapy. While macroscopic hemorrhage is relatively rare (<1%), micro-hemorrhagic change is present on haem-sensitive sequences in most cases.² A subgroup of

hypervascular CPA schwannomas in younger patients are characterized by prominent vascular flow voids.³ Additional imaging features include dural enhancement (25%) and adjacent parenchymal edema (40%). Although hydrocephalus most frequently relates to fourth ventricular obstruction, there may also be a communicating hydrocephalus due to the schwannoma secreting protein into the cerebrospinal fluid. A focal T2w hyperintensity in the dorsal brain stem is very occasionally observed and is likely related to degeneration of the vestibular nucleus (**Fig. 2**).⁴

Bilateral VSs are present in 5% of patients and are emblematic of neurofibromatosis type 2 (NF2). NF2 is an autosomal dominant tumor predisposition condition with birth incidence of 1 in 25 to 33,000. Patients with mosaic NF2 may present with unilateral VS, but there will usually be clinical features or a family history of NF2. NF2 VSs are characterized by a younger age at presentation, more rapid growth, and intracochlear extension. NF2 is also associated with other cranial nerve schwannomas and meningiomas with the potential for collision tumors in the posterior fossa. Whole neuro-axis imaging is appropriate as 75% of patients will also develop schwannomas and ependymomas within the spine (**Fig. 3**).

VSs comprise 60% to 70% of CPA tumors in major surgical series. The other most frequent primary extra-axial tumors of the CPA are meningiomas (10%–15%) and epidermoids (5%).⁵ The distinguishing imaging features of these lesions are demonstrated in **Table 2** (**Figs. 4–6**). CPA meningiomas are important to distinguish from VSs preoperatively because they demonstrate a different relationship to neurovascular structures, variable histology, and potential invasiveness, which therefore influences the surgical approach. Non-VSs in the CPA may be derived from V, VII, IX, X, XI, and XII nerves and may mimic VS both radiologically and clinically. Other primary lesions of the CPA include arachnoid cysts, metastases, developmental lesions (eg, lipoma, dermoid), and vascular lesions (eg, hemangiomas, arteriovenous

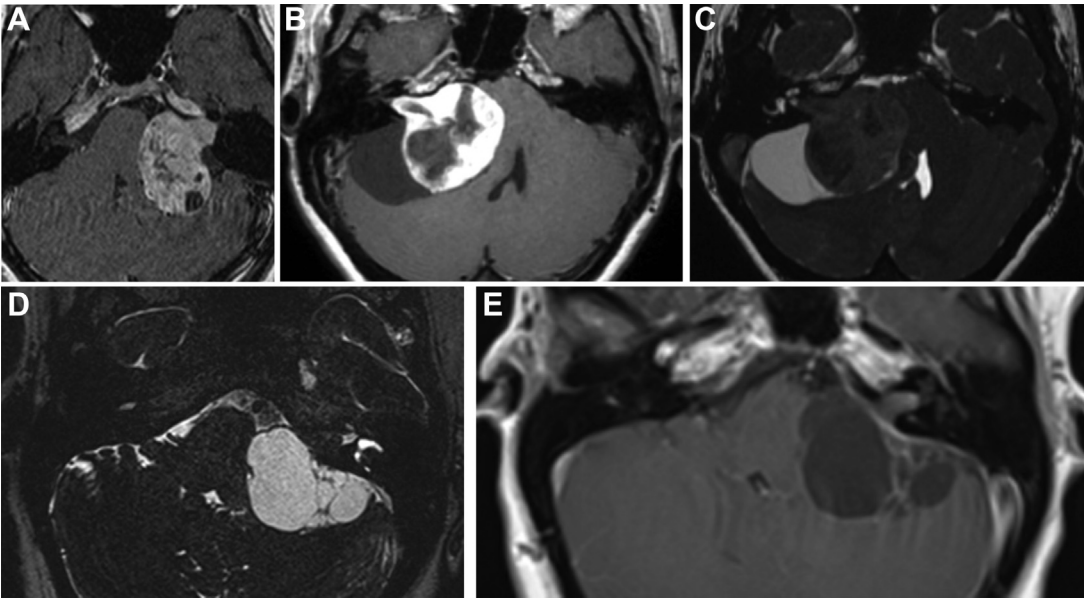


Fig. 1. Appearances of the CPA cystic schwannomas. (A) Post-gadolinium T1w axial image demonstrating intratumoral cysts while (B) and (C) post-gadolinium T1w axial and CISS axial images demonstrate a peritumoral cyst. Thin-walled cysts as shown in (D) CISS axial and (E) post-gadolinium T1w images are associated with increased risk of operative neurovascular injury.

malformations, aneurysms) (**Box 1, Table 3**). Secondary leptomeningeal involvement by metastases (eg, lung, breast, melanoma, lymphoma) and inflammatory lesions (eg, sarcoid, tuberculosis) should also be considered and will require correlation with clinical history and cerebro-spinal fluid (CSF) analysis to aid the diagnosis. It should be noted that less than 1% of patients with VS present with facial nerve dysfunction, so such a clinical presentation should raise the possibility of an alternative diagnosis such as VII nerve schwannoma, vascular lesion, or aggressive leptomeningeal disease. It should also be remembered that bilateral lesions are rare in the absence of an NF2 diagnosis, so such a finding should prompt the radiologist to consider other causes such as leptomeningeal disease.

When analyzing a CPA mass, it should be ensured that it is not arising from adjacent structures. Exophytic intra-axial lesions extending from the brain stem and cerebellum (eg, medulloblastoma in children and metastasis or hemangioblastoma in adults) or from the fourth ventricle (eg, ependymoma, choroid plexus tumor) may present as a CPA mass lesion. It should be noted that primary CPA tumors are rare in young patients, and so an exophytic intra-axial lesion should always be considered in this setting. Similarly, masses arising from the petrous apex (eg, metastasis or chondrosarcoma) or the jugular foramen (eg,

paraganglioma) may be mistaken for a CPA VS unless traced to their point of origin.

IMAGING APPEARANCES AND DIFFERENTIAL DIAGNOSIS OF THE INTERNAL AUDITORY MEATUS VESTIBULAR SCHWANNOMA

The origin of VSs is controversial, and they have been described as either arising from both the nerve sheath near the Obersteiner-Redlich zone (at the transition from central glial to Schwann cells) and from Scarpa's ganglion. The most distal part of the VIII transition zone extends over 4-5 mm which may explain the variable location of small IAM VSs. IAM schwannomas are usually hypointense to CSF on 3D FSE or GRE sequences and are described as having a nodular or fusiform morphology. Approximately 85% of VSs arise from the inferior vestibular nerve with a smaller proportion arising from the superior vestibular nerve (9%) or the cochlear nerve (6%).⁶ Dumbbell-shaped schwannomas may extend from the IAM into the cochlea (trans-modiolar), vestibule (trans-macular), or middle ear (trans-otic) (**Fig. 7**).

The differential diagnosis of IAM VSs is similar to that of those centered in the CPA (see **Box 1, Table 3**) although there are differences in the frequency of each pathology (**Fig. 8**). First, an IAM lesion is statistically even more likely to represent a VS, with schwannomas comprising over 90% of



Fig. 2. Variant appearances of the CPA vestibular schwannoma. (A) T2* axial image demonstrates VS microhemorrhage. (B) Post-gadolinium T1w axial image demonstrates a cystic VS with a dural tail (*arrow*). (C) T2 CISS image reveals a focus of T2 high signal in the region of the vestibular nucleus (*arrow*). (D) T2w axial image depicts a VS with adjacent edema while associated hydrocephalus is shown on (E) coronal post-gadolinium T1w imaging. Hypervascularity as shown on an (F) T2 CISS image and macrohemorrhage illustrated on a (G) T2w image are further variant imaging features.

all IAM tumors. A recent study of IAM lesions which were felt to be clinically and radiologically consistent with VS, demonstrated that alternative pathology was discovered at surgery in only 2.5% of cases.⁷ Second, inflammatory and neoplastic leptomeningeal disease rather more frequently presents as an IAM lesion than is the case with CPA masses. When considering inflammatory and malignant disease of the IAM, it is always important to search for an origin in the lateral petrous bone, either from inflammatory middle ear disease or from facial nerve perineural spread.

The radiologist should be particularly alert to the possibility of alternative pathologies when there is a clinical history of systemic malignancy or immunosuppression, when symptoms are rapidly

progressing, and when they include facial nerve palsy. Imaging features suggestive of another diagnosis include a linear pattern of gadolinium enhancement, bilateral lesions, and extension of disease laterally to the facial nerve or middle ear. Note that physiologic vascular gadolinium enhancement at the fundus of the IAM, normal expansion of Scarpa's ganglion, or an intrameatal vascular loop may all mimic a small VS.

CLINICAL PRESENTATION OF VESTIBULAR SCHWANNOMA AND INDICATIONS FOR DIAGNOSTIC IMAGING

Audiological symptoms are present in almost all patients with VSs. Sensorineural hearing loss is

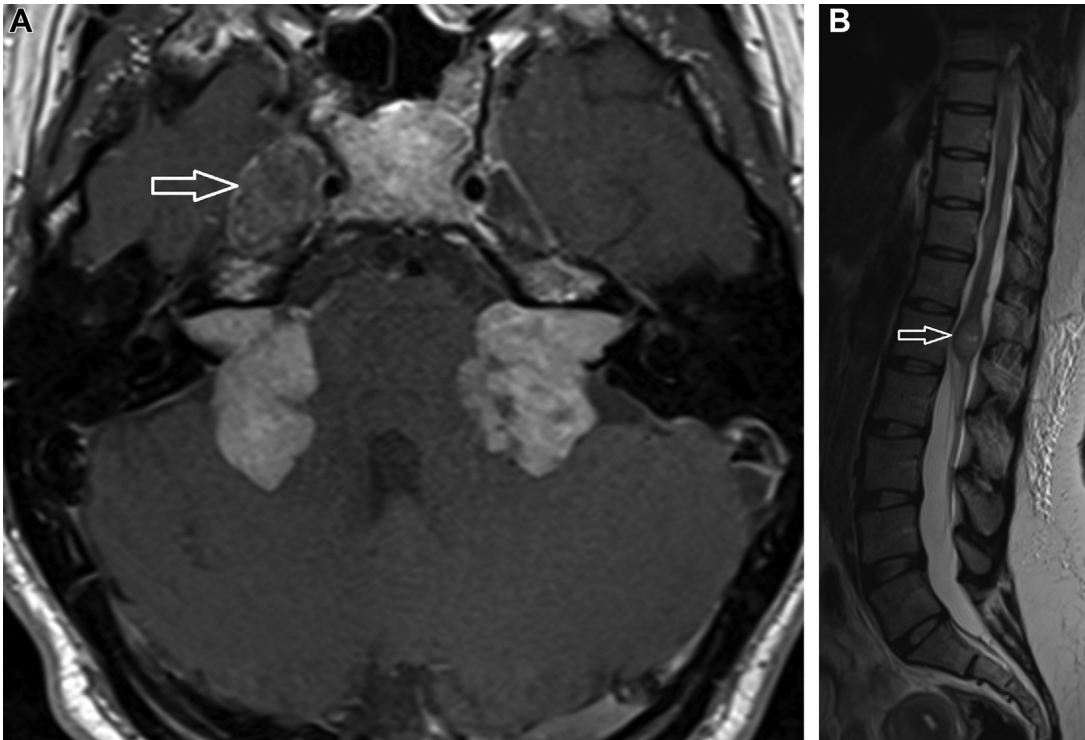


Fig. 3. Vestibular schwannomas in the context of NF2. (A) Post-gadolinium T1w illustrates bilateral vestibular schwannomas with a right trigeminal schwannoma (*arrow*) while a (B) T2w sagittal image demonstrates associated spinal cord ependymomas (largest indicated by *arrow*).

the most common symptom leading to diagnosis (95%), and this is of sudden onset in 5% of cases. Most patients also experience asymmetric tinnitus, although it is rare (<1%) for it to be an isolated presenting symptom.⁸ Larger tumors will be more likely to present with symptoms related to compression of posterior fossa structures, with the possibility of unsteadiness, true vertigo, facial pain, and numbness. Incidental diagnosis after MRI for unrelated clinical presentations may now represent up to 10% of the VSs detected.

Diagnostic imaging is required to detect VSs in patients with unilateral or asymmetric audiological symptoms or other relevant localizing symptoms and signs. In addition, MRI is indicated in the setting of asymmetric hearing loss on pure tone audiometry. There are no standardized criteria as to the required interaural audiometric threshold asymmetry to prompt imaging. The optimal choice of audiometric criteria will depend on whether the aim is to achieve maximum sensitivity (with increased numbers of MRI studies) or maximum positive predictive value. The recent National Institute of Clinical Excellence guidelines state that patients with ≥ 15 dB interaural difference in thresholds at more than two adjacent frequencies should undergo MRI; however, the audiometric

criteria remain controversial,⁹ and there are no prospective studies available. The most comprehensive criteria for imaging would be weighted for multiple factors (eg, age, history of noise exposure, other audio vestibular symptoms) and are likely to be clarified with machine learning algorithms.¹⁰

IMAGING PROTOCOLS FOR THE DETECTION OF VESTIBULAR SCHWANNOMA AND DIAGNOSTIC YIELD

Three-dimensional T2w FSE or GRE sequences have proven to be more cost-effective than gadolinium-enhanced T1w imaging¹¹ for the detection of VSs. However, there is potential for <2-mm IAM lesions, labyrinthine lesions, and leptomeningeal disease to go undetected with this approach, so there should be a low threshold to perform gadolinium-enhanced T1w imaging for equivocal cases and, in particular, clinical settings such as immunocompromised or malignancy. It should also be remembered that alternative causes for audiological symptoms (eg, inner ear pathologies) should be sought because they are detected on MRI with a similar frequency to that of VSs.¹²

Table 2
Differential diagnosis: imaging features of the three principle CPA masses

Tumour	Origin	Morphology/Location	CT Features	T1w/T2w Intensity	Enhancement	Other Sequences	Lateral to IAM Fundus
Vestibular schwannoma	Schwann cells of nerve sheath and usually inferior vestibular nerve	Centered on an expanded porus acusticus. Spherical with acute angle to dorsal petrous bone.	Variable density but principally isodense. Calcification rare.	T1 iso-intense/T2 hyper-intense	Moderate and heterogeneous with cystic non-enhancement in larger lesions	Microhemorrhage on heme-sensitive sequences. Myoinositol peak on spectroscopy	May be intralabyrinthine extension and note any extension along facial nerve canal to suggest facial schwannoma mimic.
Meningioma	Arachnoid meningoepithelial cells of CPA (or rarely IAM)	Centered on posterior petrous bone with hemispheric broad base, asymmetric to porus acusticus. Rugged medial contour.	Usually (70%) hyperdense with possible hyperostosis and calcification (20%). No enlargement of porus acusticus	Variable signal but maybe T1 iso-intense/T2 iso-intense to cortex	Moderate homogenous enhancement with dural tail	Decreased ADC compared to VS but overlapping. Increased CBV compared to VS. Alanine peak on spectroscopy	Perilymphatic signal is preserved on CISS sequences when IAM extension
Epidermoid	Inclusions of ectodermal cell rests	Insinuating within the basal cisterns, with a fine irregular surface. Extending to contralateral CPA and superior to tentorium.	Usually CSF density with occasional peripheral calcification and molding of petrous bone	Usually iso-intense to CSF signal with rare T1 hyper-intense, T2 hypo-intense "white epidermoids"	Non-enhancing	Increased DWI, FLAIR signal relative to CSF distinguishes from arachnoid cyst. Well defined with 3D T2w sequences. Lactate peak on spectroscopy	

Abbreviations: ADC, apparent diffusion co-efficient; DWI, diffusion weighted imaging; FLAIR, fluid-attenuated inversion recovery.

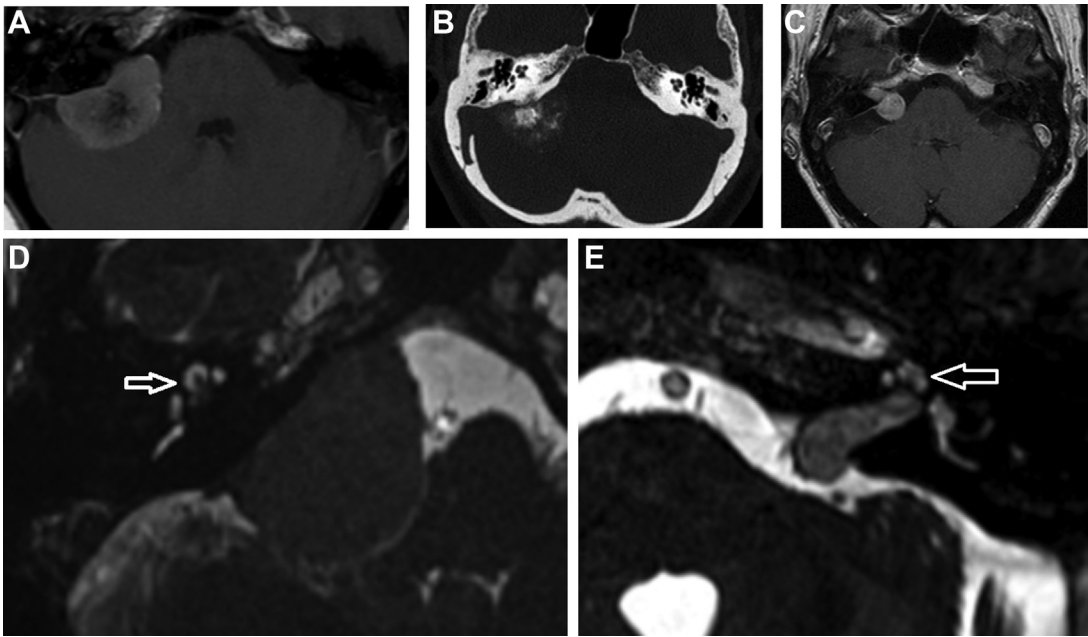


Fig. 4. CPA meningioma imaging differential diagnosis. (A) Post-gadolinium T1w axial image of a meningioma reveals the broad base along the posterior petrous pyramid and its location eccentric to the IAM with (B) calcification on CT. (C) Post-gadolinium T1w axial images showing meningeoma with intracanalicular extension. Note how perilymphatic signal is preserved on CISS sequences when there is IAM extension from a (D) meningioma, whereas it may be reduced with an (E) VS (arrows).

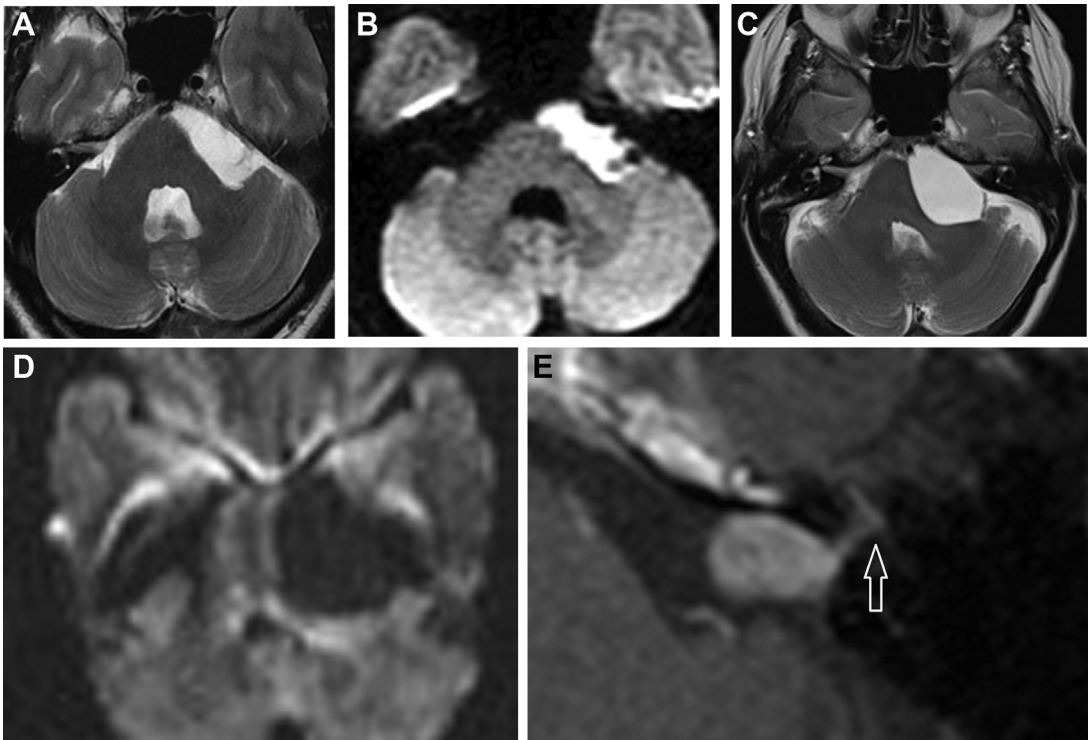


Fig. 5. Imaging differential diagnosis of CPA mass. (A) T2w axial and (B) DWI depict a CPA epidermoid and characterized by increased DWI as compared to (C) T2w axial and (D) DWI images of a CPA arachnoid cyst. (E) Post-gadolinium T1w axial image identifies a facial schwannoma by its tail of enhancement in the line of the nerve (arrow).

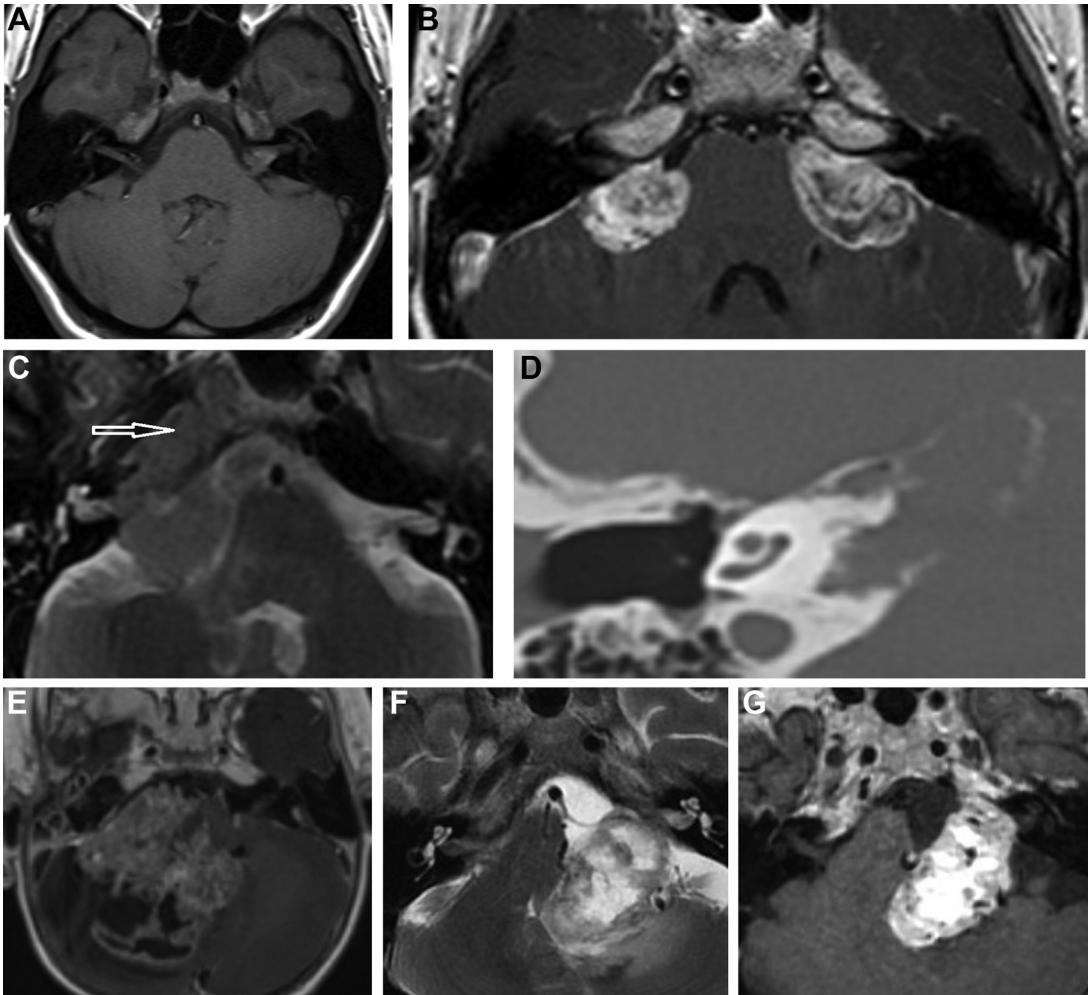


Fig. 6. Pitfalls in the characterization of a CPA mass. (A) T1w axial image with patchy T1 high signal due to a low flow venous malformation. (B) Post-gadolinium T1w axial image with bilateral CPA lesions and additional meningeal enhancement secondary to leptomeningeal atypical rhabdoid teratoid tumor. (C) T2w axial image with CPA exophytic extension from a petrous apex (*arrow*) metastasis with the bony changes confirmed in the (D) CT study. (E) Post-gadolinium T1w axial image demonstrates CPA exophytic extension from an intra-axial GBM. (F) and (G) T2w and post-gadolinium T1w axial images demonstrating a hemangioblastoma extending exophytically into the CPA from a pial attachment to the lateral pons.

As audio-vestibular symptoms are common, and audiometric selection criteria are now more liberally applied, the imaging for detection of VSs now represents a significant burden for many imaging departments. It has been estimated that 20% of patients referred to ear, nose, and throat secondary care will satisfy audiometric criteria for MRI.¹² As MRI has become more available, there has been a trend toward lower diagnostic yield for the detection of VS. Although varying in selection criteria, studies of (>300 patient) populations with asymmetric symptoms or audiometry over the past 20 years have demonstrated schwannomas in 1.4%-9.2% of cases.¹²⁻¹⁴ The

increasing use of MRI has also contributed to the rising incidence of VS, which has been documented on several national databases. This is associated with a reduction in mean tumor size at detection from 26 mm to 7 mm.¹⁵

DIFFERENT MANAGEMENT OPTIONS FOR VESTIBULAR SCHWANNOMA AND THE ROLE OF IMAGING

The three principle approaches available for the management of VSs are a “wait and scan” conservative approach, surgery, and radiotherapy. Less frequently, medical therapy may be considered;

Box 1**Radiological differential diagnosis of a CPA or IAM lesion**

Vestibular schwannoma

Meningioma

Non-vestibular schwannoma

Developmental: epidermoid/lipoma/hamartoma

Vascular: aneurysm/arteriovenous malformation/venous malformation

Leptomeningeal disease: infection/inflammation/neoplasia (lymphoma or metastasis)

however, this is principally in the setting of NF2. The therapeutic decision-making is multifactorial with clinical (eg, age and symptoms) as well as imaging findings (size and growth) being considered. In addition to its influence on the choice of management option, imaging plays a key role in the planning of interventions and posttreatment monitoring.

Conservative Wait and Scan

As most VSs are small and slow growing, “wait and scan” protocols are playing an increasing role, particularly in older patients. Measurement techniques, appropriate timing of imaging, optimal imaging protocols, and definition of tumor growth will be discussed.

Tumor Measurements

Consistent measurement techniques and reliable measurements of tumor size are important to assess the tumor size and growth rate. There is considerable variation in how measurements are performed, in both clinical practice and the research setting (Fig. 9).¹⁶ Standardized methods of tumor measurement have been promoted by organizations, although none have been uniformly adopted.¹⁷ The largest extrameatal dimension was deemed the key measurement at a recent consensus meeting, and it forms the basis for most tumor staging classifications (Table 4). Although these single linear measurements have been widely adopted, it is known that they are prone to measurement error and that the reliability

Table 3**Pearls and pitfalls in the imaging assessment of a CPA or IAM lesion**

Imaging Review Area	Why?
Evidence of cysts	Propensity for tumors to grow rapidly and impacts on effectiveness and morbidity of surgery/radiotherapy
Extension of enhancement to other cranial nerve foramina and canals	Consider a nonvestibular schwannoma which influences surgical approach
Interface with petrous apex, jugular foramen and brain	Ensure it is a primary CPA lesion rather than an exophytic extension from adjacent structures
Look for contiguity with disease in the lateral petrous bone	Acute and chronic inflammatory middle ear disease and facial nerve perineural spread can demonstrate IAM extension
Review pre-gadolinium T1w sequence	Do not misinterpret T1w hyperintensity (eg, lipoma) as gadolinium enhancement
Note if extrameatal component of the mass is eccentric to IAM and forming an obtuse angle with petrous bone	Consider a meningioma with intracanalicular component which effects surgical approach
Look for excessive flow voids and vascularity	Highlight hypervascular VS variant to surgeon or consider alternative vascular pathology
Reformat volumetric sequence to standard plane	Ensures consistent measurement technique for assessment of tumor growth
Identify other leptomeningeal lesions or bilateral lesions in absence of NF2 diagnosis	Suggest alternative diagnosis of inflammatory/infective/neoplastic leptomeningeal disease

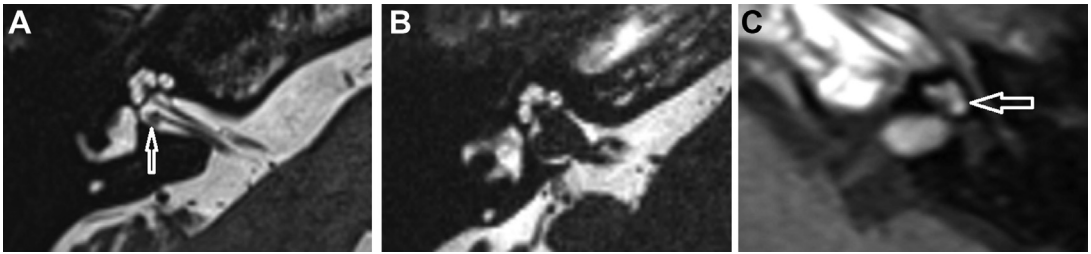


Fig. 7. IAM vestibular schwannoma appearances. CISS axial images revealing an (A) small nodular inferior vestibular nerve schwannoma (arrow) and a (B) fusiform schwannoma extending along the inferior vestibular and cochlear nerves. (C) Post-gadolinium T1w image reveals transmodiolar extension (arrow).

and agreement can be improved by combining multiple linear dimensions or evaluating area dimensions.¹⁸

Further precision may be achieved by applying volumetric analysis, which is recognized to have the optimum ability to define growth between serial scans when accounting for measurement error. Although algorithms are rapidly evolving, there are currently challenges to the routine use of volume analysis in clinical practice. Completely automatic segmentation is generally not possible, particularly in view of the difficulties posed by adjacent dural and venous enhancement, necrosis, and collisions tumors.¹⁹ The ability to accurately segment the tumor is also dependent on the imaging acquisition, with the contrast relative to adjacent tissues being suboptimal when T2w

sequences are used, and the measurement error increasing to greater than 10% when less than five sections pass through the tumor.²⁰ With the exception of the monitoring of medical therapy, linear dimensions currently remain adequate for most clinical decision-making and are able to predict volumetric progression with 70% to 90% accuracy.^{21,22} Artificial intelligence methods have been applied to fully automated segmentation and volumetric assessment so may have a future role in VS surveillance.

Natural history of VSs as a guide to optimizing “wait and scan” imaging follow-up interval and duration

In order to guide follow-up imaging protocols for the conservative management of VSs, it is

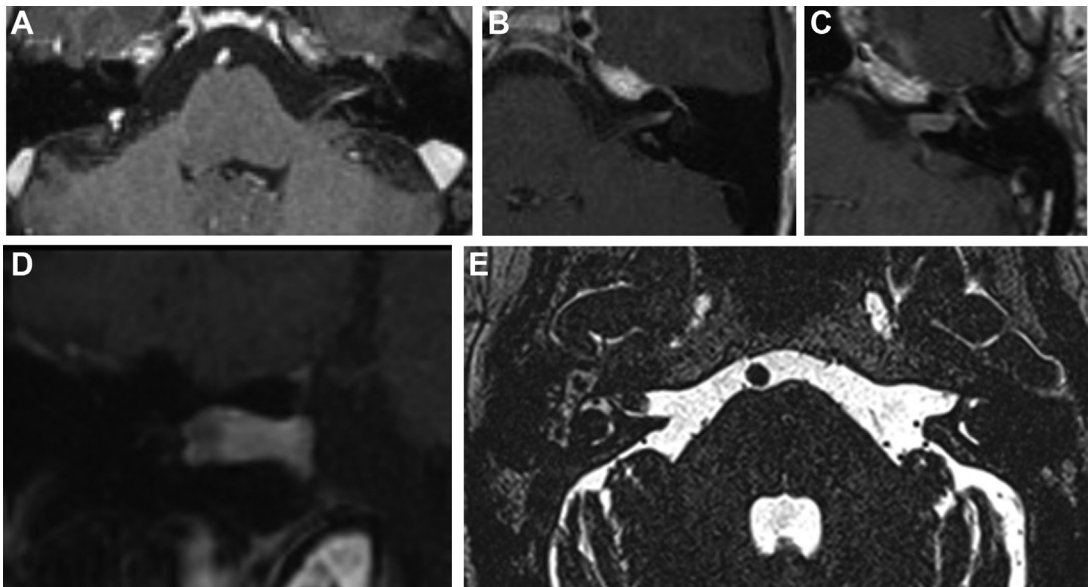


Fig. 8. Differential diagnosis of IAM lesion. (A) Post-gadolinium T1w axial images demonstrates linear IAM enhancement related to (A) neurosarcoïd, (B) Lyme disease, and (C) adenoid cystic carcinoma perineural spread. (B) and (C) demonstrate extension in the *line* of the facial nerve. (D) Post-gadolinium T1w image also demonstrates an intra-canalicular meningioma, whereas (E) CISS imaging demonstrates a focal abnormality in the IAM secondary to tympanomastoid inflammation.

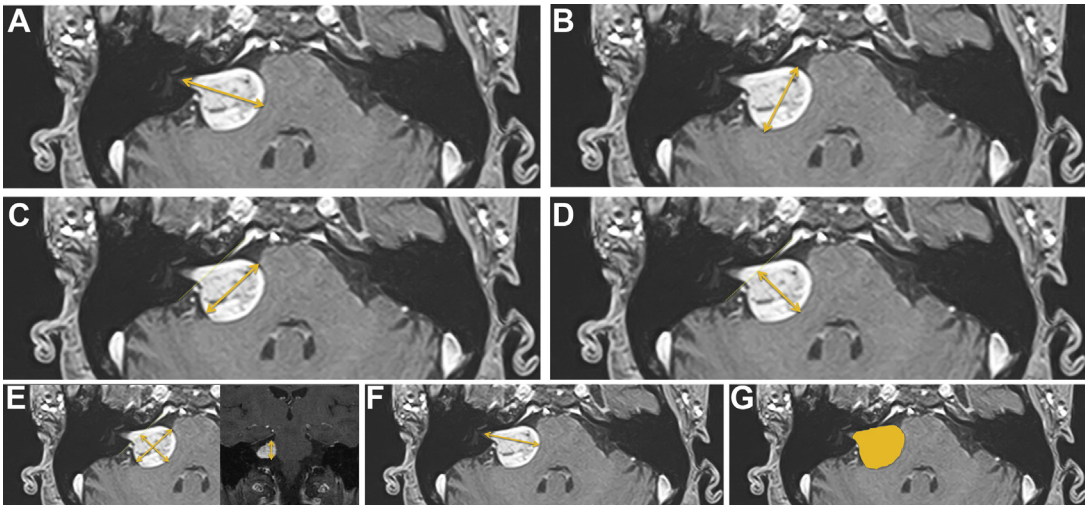


Fig. 9. Different VS measuring methods (indicated by double arrows). Post-gadolinium T1w axial images demonstrate previously described measurements for the documentation of VS size: (A) Longest axial dimension, (B) maximum extrameatal dimension, (C) maximum extrameatal dimension parallel to posterior petrous pyramid, (D) maximum extrameatal dimension perpendicular to posterior petrous pyramid, (E) combination of dimensions perpendicular and parallel to posterior petrous pyramid with a craniocaudal dimension (AAOHNS method), (F) longest axial dimension in the *line* of the IAM and (G) area dimension.

important to have some knowledge of their natural history. CPA VSs grow at a mean rate of 1 to 2 mm/y and approximately 33% of tumor volume/y, but this is variable, with growth varying over time. Larger tumors at presentation and cystic tumors have greater propensity to growth with approximately 10% of tumors demonstrating rapid growth of greater than 1 cm/y. Between 20% and 50% of tumors undergo a degree of growth which requires intervention, and this usually occurs in the first 2 to 3 years after diagnosis. The longer the VS has been observed to be stable, the lower the risk of subsequent growth is, but in 5% to 10% of patients, it should be noted that the growth first occurs after 5 years.^{23,24} In 5% to 20% of tumors, it

has been observed that tumors will undergo spontaneous regression.

Therefore, as most significant growth occurs in the first 2 to 3 years after presentation, this should represent the most intensive period of monitoring; however, longer term follow-up is also required to detect growth. There is no standardized protocol, although most guidelines propose yearly imaging for 3 to 5 years before increasing the duration between MRI studies in the context of a stable tumor.

MRI sequences for “wait and scan” imaging follow-up

A series of studies have now shown that 3D T2w MRI sequences are equivalent to gadolinium-

Table 4
Staging systems for vestibular schwannoma

Tumor Size	House	Koos		Samii: Tumor Extent
Intracanalicular	Intracanalicular	Grade I	T1	Isolated to IAM
≤10 mm	Grade 1 (small)	Grade II	T2	Minor extrameatal extension
≤15 mm	Grade 2 (medium)		T3a	Extrameatal tumor without brain stem contact
≤20 mm				
≤30 mm	Grade 3 (moderately large)	Grade III	T3b	Tumor contacting brainstem without compression
≤40 mm	Grade 4 (large)	Grade IV	T4a	Tumor compressing brainstem
>40 mm	Grade 5 (giant)		T4b	Tumor severely compressing brainstem and fourth ventricle

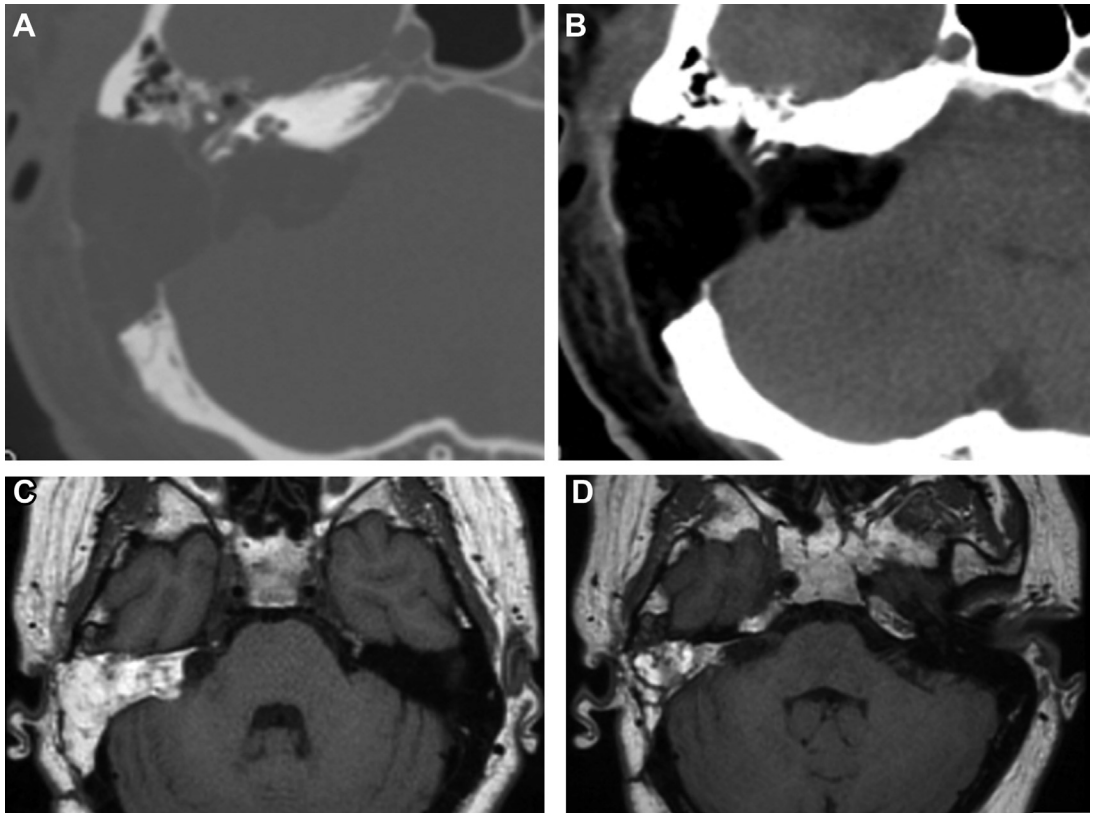


Fig. 10. Translabrynthine postsurgical appearances. CT axial with (A) bone and (B) soft-tissue windows indicating the expected petro-mastoid bony defect and fat graft to the subtotal petrosectomy cavity. T1w axial images (C) 3 months after surgery and (D) 4 years after surgery demonstrate interval involution of the fat graft.

enhanced imaging in terms of their ability to predict tumor progression, with less than 0.5 mm mean difference in measurements obtained between the two different types of sequences. Although guidelines indicate that either MRI approach is acceptable, it is likely that 3D T2w sequences will be adopted because of concerns about the long-term safety of multiple administrations of gadolinium-based agents.²⁵

Tumor growth and failure of “wait and scan” conservative management

Demonstration of tumor growth is critical for deciding whether a “wait and scan” conservative approach has failed and whether specific interventions are required, so “significant” growth needs to be defined. A recent position statement²⁶ specified growth as greater than 2 mm linear measurement compared with the index study, or alternatively a 1.2 mm³ or greater than 20% increase in volume. The concept of growth rate, rather than absolute growth, was also introduced as a feature requiring a change in management strategy.²⁶ As the ability to define growth will depend on the imaging and

measurement technique, it could be argued that the smallest detectable difference between serial imaging studies should be calculated locally, on the basis of departmental agreement parameters. It is likely that future approaches will include advanced imaging parameters and genetic alterations to help predict tumor behavior and a propensity for growth.²⁷

It should be appreciated that growth is not the only factor to indicate whether surgery or radiotherapy is to be offered. Absolute tumor size is important, and it is noted that larger tumors are associated with younger patients, possibly reflecting a more aggressive tumor biology. In addition, patient preference, age, comorbidities, and clinical features (eg, hearing loss) are all considered by the multidisciplinary team.

IMAGING IN THE SURGICAL MANAGEMENT OF VESTIBULAR SCHWANNOMAS *Surgical Approaches for Vestibular Schwannoma*

Surgery may be required to address large or growing CPA VSs, and the main surgical options are the

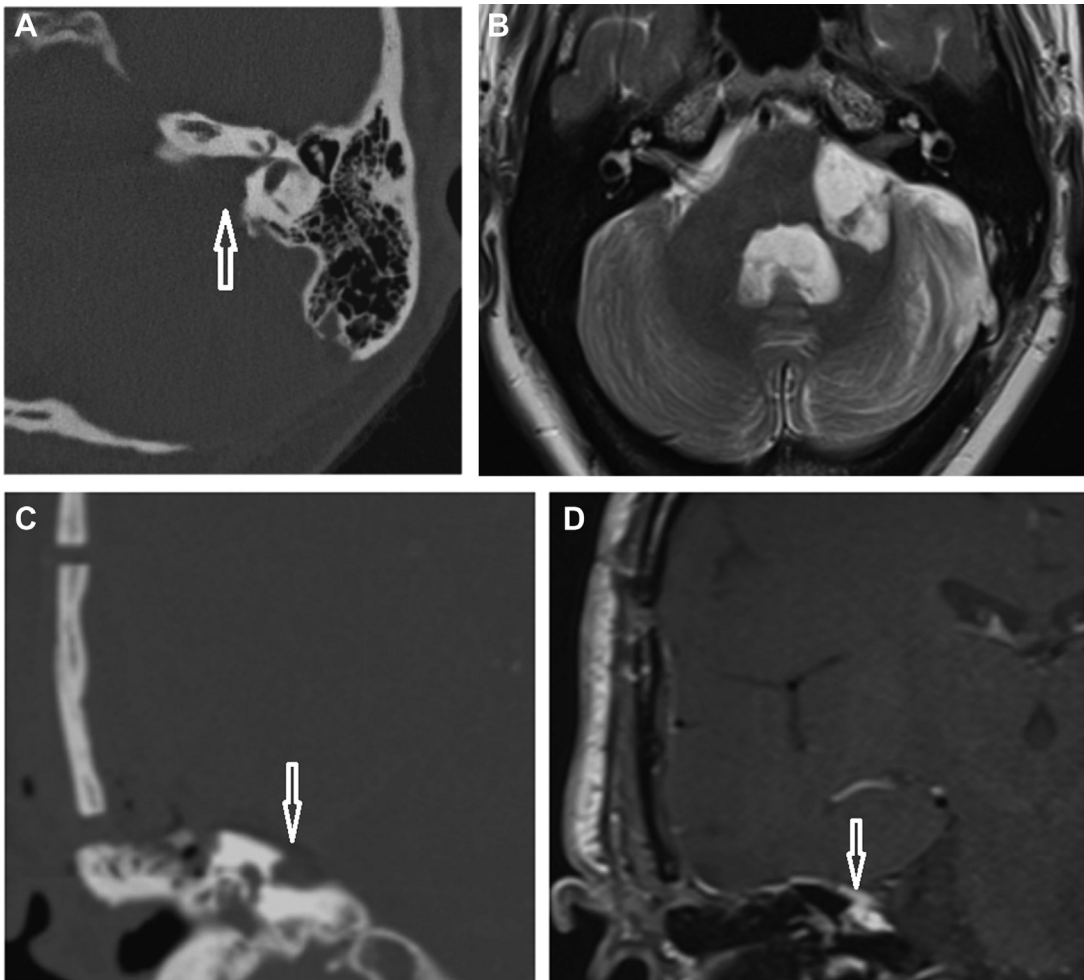


Fig. 11. Retrosigmoid and middle cranial fossa approach postsurgical appearances. Post retrosigmoid approach appearances: (A) axial CT demonstrates the expected occipito-mastoid and posterior porus acusticus (*arrow*) bony defects while the (B) T2w axial image reveals left cerebellar encephalomalacic changes and widening of the left CPA. Post-middle cranial fossa approach appearances: (C) Coronal CT demonstrates the temporal craniotomy and defect within the superior IAM (*arrow*) with (D) T1w coronal demonstrating a fat graft at the site of the IAM defect (*arrow*).

retro-sigmoid (suboccipital) or translabyrinthine surgical approaches (Figs. 10 and 11). The middle cranial fossa approach is less frequently used to remove intracanalicular tumors, or those with small extrameatal components. There is potential for hearing preservation with translabyrinthine and middle cranial fossa surgical routes. A summary of the surgical steps is listed in Table 5, together with the advantages and role of each operative approach.^{5,28,29} Surgery is generally preferred to radiotherapy in younger patients, patients presenting with trigeminal neuralgia, and when there are significant cystic components. A surgical approach is also favored for larger tumors because of concerns about compressive ischemia of the facial nerve or brainstem due to tumor swelling with radiotherapy.

Preoperative imaging assessment

A check list for a radiological review of preoperative imaging is proposed in Table 6 (Fig. 12), with the most relevant preoperative imaging features depending on the intended surgical approach. The key observations which should be identified include

- Venous variants which may encroach on the surgical pathway;
- Facial nerve location anterior to the VS which may lead to inadvertent injury;
- Extension of petro-mastoid air cells which will require sealing to prevent CSF leak;
- Lateral extension of tumor within the IAM which may limit resection or result in increased morbidity;

Table 5
Surgical approaches for vestibular schwannoma resection

Surgical Approach	Stages of Surgical Technique	Advantages	Main Role	Risks/Potential Postoperative Complications
Middle cranial fossa	<ul style="list-style-type: none"> • Preauricular approach • Temporal craniotomy • Temporal lobe retraction • Medio-lateral decompression of the superior IAM and open dura • Remove tumor • Graft to dural defect 	<ul style="list-style-type: none"> • Good access to fundus • May preserve hearing 	Intracanalicular lesions or limited extrameatal component	<ul style="list-style-type: none"> • Slight increased risk of facial nerve injury • Temporal lobe seizures from temporal lobe retraction
Retrosigmoid (suboccipital)	<ul style="list-style-type: none"> • Postauricular approach • Retrosigmoid craniectomy inferior to transverse sinus • Seal any mastoid air cells • Dura incised and arachnoid opened • Drain CSF and retract cerebellum • Remove tumor • Facial nerve usually identified later at brainstem after debulking • Access IAM component by medio-lateral decompression posterior two-third (to avoid labyrinth) IAM 	<ul style="list-style-type: none"> • Wide exposure to CPA • <i>Compared to trans-labyrinthine:</i> • May preserve hearing 	CPA lesions of any size with view to preserving hearing Cannot preserve hearing if far lateral IAM extension	<ul style="list-style-type: none"> • CSF leak into petro-mastoid air cells • Aseptic meningitis • Cerebellar atrophy • Early postoperative headaches

Trans-labyrinthine

- Postauricular approach
- Subtotal petrosectomy (anterior to sigmoid sinus)
- (Partial) labyrinthectomy
- Sigmoid sinus can be retracted and jugular bulb displaced inferiorly
- 270° removal of IAM
- Access CPA dorsal to porus acusticus
- Tumor removal
- Facial nerve identified early in IAM and lateral at brainstem
- Packing of middle ear
- Fat graft to subtotal petrosectomy
- Wide exposure to CPA
- *Compared to retrosigmoid:*
- Extradural bone drilling
- Identification of facial nerve early in IAM
- Less cerebellar retraction
- CPA lesions of any size in patients without serviceable hearing or poor hearing prognosis
- Can address intralabyrinthine extension
- Hearing eliminated
- Complications of fat harvesting
- Sigmoid sinus and jugular bulb thrombosis from retraction

Table 6
Preoperative and postoperative imaging assessment

Surgical Approach	What the Surgeon Needs to Know? Preoperative Imaging Features of Surgical and Clinical Importance	What the Radiologist Should Expect to Find? Postoperative Imaging Features
Middle cranial fossa	<ul style="list-style-type: none"> • Far lateral extension below crista falciformis/cochlear aperture (reduces possibility of hearing preservation) 	<ul style="list-style-type: none"> • Temporal craniotomy • Fat/fascial graft to roof of IAM
Retrosigmoid (suboccipital)	<ul style="list-style-type: none"> • Extensive petromastoid pneumatization (risk of postoperative CSF leak if air cells not sealed with those adjacent to IAM particularly important) • High jugular bulb behind IAM (assess requirement for displacement and potential morbidity) • Inner ear signal change on T2w 3D/FLAIR (reduces possibility of hearing preservation) • Extension of tumor to fundus of IAM (may increase risk to facial nerve and result in fenestration when accessing lateral IAM so decreased possibility of hearing preservation) • Facial nerve location/deflection relative to tumor in CPA (increased risk of injury if surgeon unaware of rare anterior position relative to tumor) 	<ul style="list-style-type: none"> • Retrosigmoid craniectomy • Absent posterior wall IAM • Cerebellar signal change and lateral flattening • Posterior fossa extra-axial collection • Linear enhancement at IAM
Translabyrinthine	<ul style="list-style-type: none"> • Anterior sigmoid sinus and high jugular bulb (assess requirement for retraction/displacement and potential morbidity) • Presence of intact torcula (to assess risk of postoperative sigmoid sinus or jugular bulb thrombosis due to retraction) • Facial nerve location/deflection relative to tumor in CPA (increased risk of injury if surgeon unaware of rare anterior facial nerve position relative to tumor) 	<ul style="list-style-type: none"> • Subtotal petrosectomy • Triangular fat graft to cavity • May be faint enhancement at margins of graft • Fat graft involutes with time • Linear enhancement at IAM

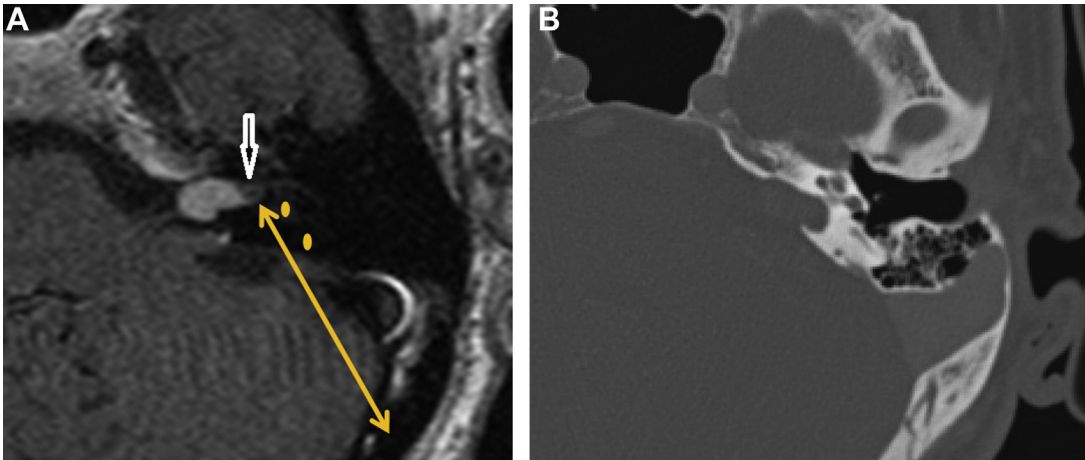


Fig. 12. Preoperative imaging assessment for VS surgery. (A) Post-gadolinium T1w image illustrates how to estimate the access to the fundus of the IAM via a retro-sigmoid route. An *arrowed line* extending from 2 cm posterior to the sigmoid sinus and medial to the line of the posterior semicircular canal (*dots*). The distance of the lesion from the fundus is also noted (*arrow*) (B) CT demonstrates a markedly anterolateral sigmoid sinus (*arrow*) which would impede access via a translabyrinthine route.

- Inner ear CISS or 3D FLAIR signal abnormality which indicates a poorer hearing outcome.^{28–30}

There has been increased interest in the potential of MRI techniques to demonstrate the position of the cisternal facial nerve with respect to the tumor. Despite microscopic inspection and intraoperative monitoring, there remains only 50% to 70% preservation of facial nerve function when operating on tumors greater than 2 cm in size. Less than 10% of facial nerves lie along the anterior or inferior aspect of the tumor, and these are

particularly important to identify preoperatively, as they will be encountered early in the resection.³¹ As routine T2 3D sequences are less successful in demonstrating the interface with the flattened or attenuated facial nerve in the larger VS, the potential for facial nerve tractography to predict facial nerve position has been explored (**Fig. 13**). Technical aspects are reviewed in more detail later in this issue. A recent systematic review and pooled analysis of 14 studies (n = 234) has demonstrated the facial nerve to be verified and concordant with surgical findings in 87%.³² Prospective blinded studies using probabilistic tractography are ongoing, and the potential for integration into neuro-navigation systems as well as the actual impact on preserving facial nerve function will require further evaluation.

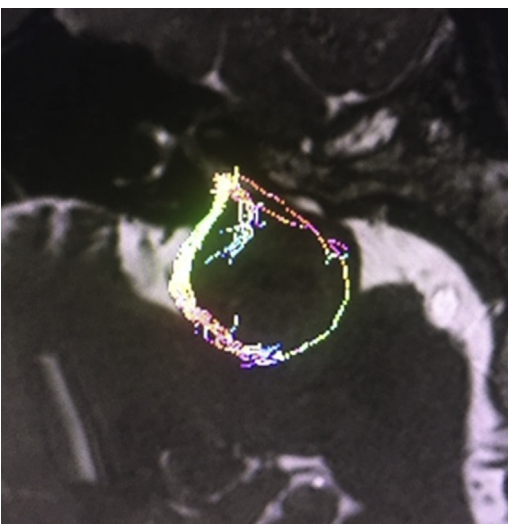


Fig. 13. Facial nerve tractography. Probabilistic DTI tractography fused with a T2 CISS image demonstrates the cochlear nerve lateral and the facial nerve medial to the vestibular schwannoma.

Postoperative Imaging Assessment

The expected postoperative MRI appearances for each of the surgical approaches are described.^{28,29} Imaging may be required to evaluate postoperative complications. Potential postoperative complications depend on the surgical approach (see **Table 6**) with imaging being required to assess those due to CSF leaks (and associated meningitis) or parenchymal injury. In the context of CSF leaks (1%–8%), CT and 3D T2w MRI may be used to identify breaches in unsealed petro-mastoid air cells, at the margins of the craniectomy or at the petrous apex, and the associated transgression of CSF (**Fig. 14**). Fat graft necrosis (1%) after translabyrinthine surgery may also predispose to CSF leak and manifests as fragmentation and fluid infiltration of the graft with potential lipid meningitis and

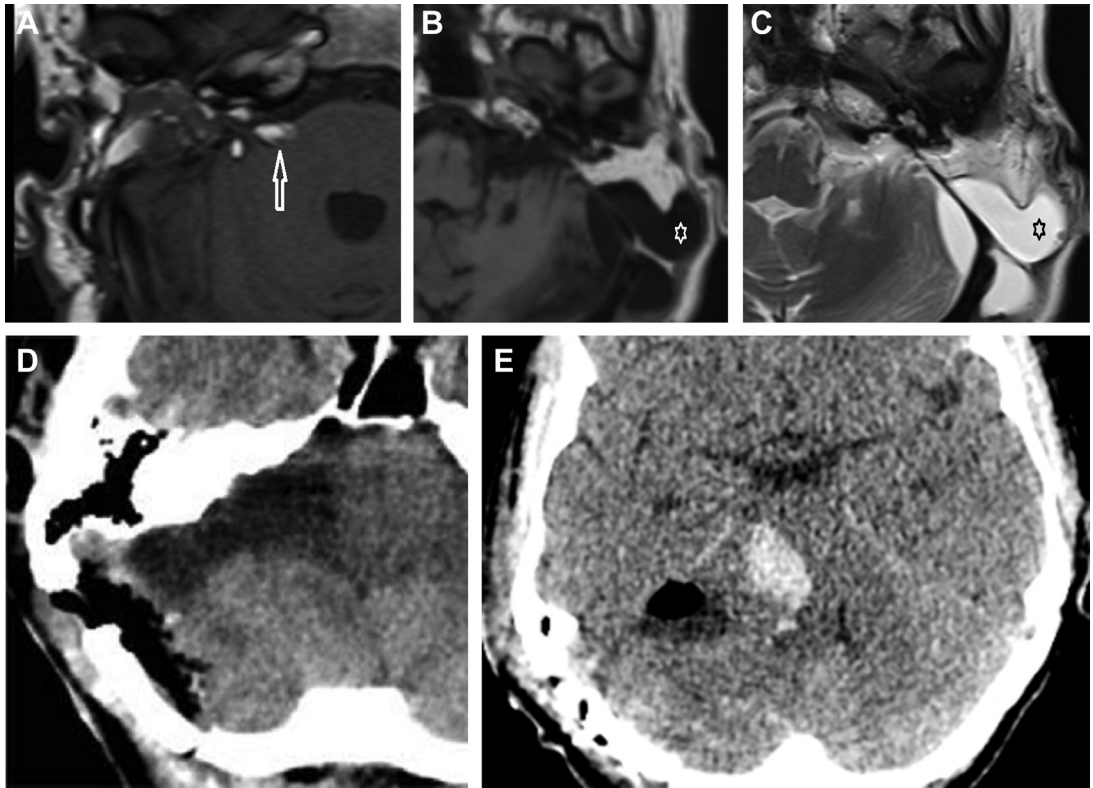


Fig. 14. Postoperative complications. (A) T1w axial demonstrates fragmentation of the fat graft with subarachnoid dissemination (*arrow*). (B) T1w post-gadolinium and (C) T2w images demonstrate fat graft necrosis with development of a pseudomeningocele (*star*). (D) and (E) CT axial images made early after surgery illustrate low-density bone wax mimicking air deep to the craniotomy (*arrow*) while acute hemorrhage is demonstrated in the surgical bed.

pseudomeningocele. Bone wax used to seal air cells or control bleeding from emissary veins should not be mistaken for intracranial air at the site of surgery or dural venous thrombus. Brain parenchymal injury may be secondary to arterial or venous ischemia, cerebritis, or retraction injury.²⁸ Postoperative hemorrhage (0.6%) may result from venous injury, with particular risk to bridging petrosal veins which are adherent to almost all large tumors. There is also a risk of ipsilateral dural venous sinus thrombosis (5%–6%).

Postoperative Imaging for Monitoring

Although a gross tumor resection (GTR) may be achieved, the goal of surgery has shifted from total resection to functional preservation. Near total resection (NTR) with less than 5% residuum, or subtotal resection (STR) with greater than 5% residuum, may be observed. In the context of a GTR, enhancement is still expected at the site of resection due to inflammation and early fibrosis. At 1 year after the surgery, it is possible to stratify the risks of recurrence according to the pattern of enhancement with linear enhancement (present in

3%) rarely progressing,³³ while there is a 6 to 16 times increased recurrence rate with nodular enhancement.³⁴ There is no standardized approach to the monitoring of GTR, but stable MRI changes are generally followed up for 3 to 5 years, depending on whether there is linear or nodular enhancement. The requirement for gadolinium enhancement for subsequent MRI in this setting is controversial.³⁵ In the context of NTR and STR, the tumor remnant is usually related to the facial nerve (**Fig. 15**). Monitoring MRI protocols for NTR and STR are similar to those for preoperative tumor; however, small tumor remnants are noted to demonstrate more indolent biological behavior.

RADIOTHERAPY FOR VESTIBULAR SCHWANNOMAS

The main alternative therapeutic intervention for VSs is radiotherapy, and this is most frequently delivered as stereotactic radiotherapy (SRT) with gamma knife or a linear accelerator. In most small- to medium-sized VSs (<3 cm), SRT is recognized as the primary treatment³⁶ and is more cost-effective. When

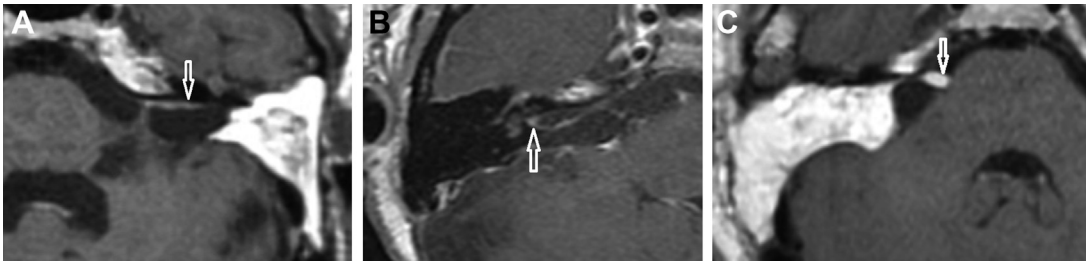


Fig. 15. Postoperative monitoring of the VS remnant. Post-gadolinium T1w axial images demonstrate (A) a linear pattern of enhancement (*arrow*) which is a normal postoperative finding and which (B) can sometimes appear more tumefactive and thicker (*arrow*), whereas (C) illustrates residual VS in the line of the facial nerve (*arrow*).

corrected for expected natural history of the tumor, there is a 75% control rate achieved with SRT³⁷ given at 11 to 13 Gy, and with similar long-term hearing outcomes.

Pretreatment MRI is principally required for the contouring of the tumor and to calculate accurate dose volume histogram. Effective tumor segmentation has now proven possible with 3D T2w imaging. Some imaging features may also prove useful in predicting outcomes, with maintained cochlear CISS signal being associated with hearing preservation,³⁸ while some texture feature parameters and lower pretreatment ADC values have correlated with response to radiotherapy.³⁹

Expected changes after SRT include a transient enlargement which occurs in 25% to 60% of patients depending on the definition.^{40,41} This should not be misinterpreted as tumor progression which is only observed in a small proportion of these enlarging tumors. Subsequent shrinkage of the tumor usually occurs within 2 years of treatment, with regression to a volume smaller than the original size occurring over 20 to 55 months.⁴⁰ The tumor swelling is often associated with reduced central enhancement (necrosis), but this does not correlate with success of treatment. Radiotherapy may also result in adjacent brain parenchymal signal change (30%) which usually resolves (**Fig. 16**). There are concerns about SRT inducing malignant transformation in schwannomas although there are

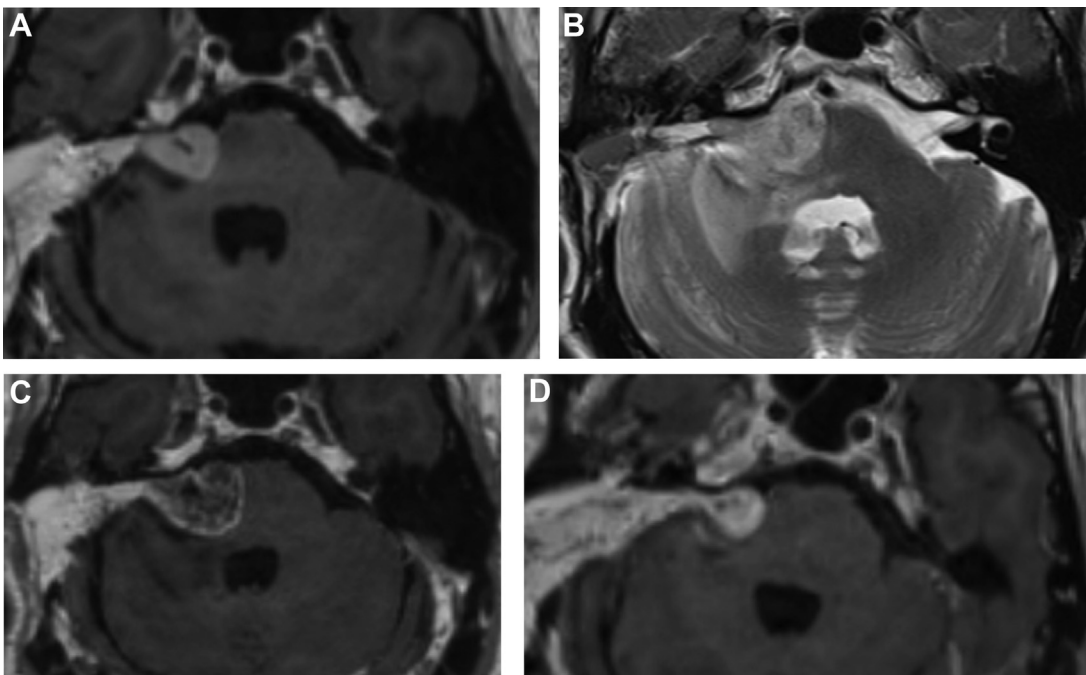


Fig. 16. Radiotherapy-related imaging changes. (A) Post-gadolinium T1w axial image demonstrates a recurrent VS after previous transabyrinthine surgery. (B) and (C) T2w and post-gadolinium T1w axial images at 6 months after SRT shows expansion ("pseudoprogression") of the VS with necrosis and adjacent parenchymal edema. At 2 years after SRT, the (D) post-gadolinium T1w image demonstrates regression of the VS.

only a few reports of definitively benign VSs becoming malignant after radiotherapy.

MEDICAL THERAPY FOR VESTIBULAR SCHWANNOMA

Bevacizumab (Avastin) is a vascular endothelial growth factor-binding antibody which has been reported to induce tumor shrinkage and improve hearing in 40% to 60% of NF2-related progressive VSs.⁴² Additional biological therapeutics such as lapatinib (an ErbB2/EGFR inhibitor) are also being evaluated. Volumetric changes in tumor size are key to defining eligibility for initiation and continuing of treatment, while also providing robust outcome tools for clinical trials. Imaging may also play a future role in the prediction of response to these medical therapies, and a series of studies have already shown promising associations between response to bevacizumab and imaging markers such as Ktrans and mean ADC.⁴³

SUMMARY

Imaging is integral to the diagnosis, treatment planning, and follow-up of VSs. MRI has led to increased detection of symptomatic and incidental small tumors,⁴⁴ which require long-term monitoring. Developments in MRI techniques and imaging analysis are continuing to strengthen the role that radiology plays in the management of the VS.

DISCLOSURE

The author acknowledges funding support from Wellcome/Engineering and Physical Sciences Research Council Centre for Medical Engineering at King's College London (WT 203148/Z/16/Z); National Institute for Health Research Biomedical Research Centre at Guy's & St Thomas' Hospitals and King's College London; Cancer Research UK National Cancer Imaging Translational Accelerator (A27066); the UK Research & Innovation London Medical Imaging and Artificial Intelligence Centre.

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