



SAPIENZA  
UNIVERSITÀ DI ROMA

Roma  
19 ott 2018

# Intralabyrinthine Schwannomas

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“La Sapienza”

Vestibular schwannomas are benign slow-growing tumors that arise from the eighth cranial nerve

Rarely, vestibular schwannomas may arise primarily within the inner ear.

These tumors have been collectively termed **intralabyrinthine schwannomas** and are now recognized as a distinct clinicopathologic disease.

Intra-labyrinthine schwannomas are benign tumours arising out of neural elements of the **terminal branches of the VIII cranial pair**, primarily affecting the vestibule, the semicircular canals, and/or the cochlea although, as they grow, they may extend into the internal auditory conduct, the pontocerebellous angle and the middle ear.

It represents a rare entity with a prevalence of 0.1% to 1% in autopsy reviews

These lesions can grow over time, often crossing into anatomically distinct portions of the inner ear, as well as into the internal auditory canal.

Before to high-resolution MRI of the inner ear, **intralabyrinthine schwannomas** was commonly diagnosed as an incidental finding during labyrinthectomy or autopsy, with the first case being reported in 1917

In 2004, Kennedy et al. proposed a classification system that was quickly adopted by the neurotologic community.

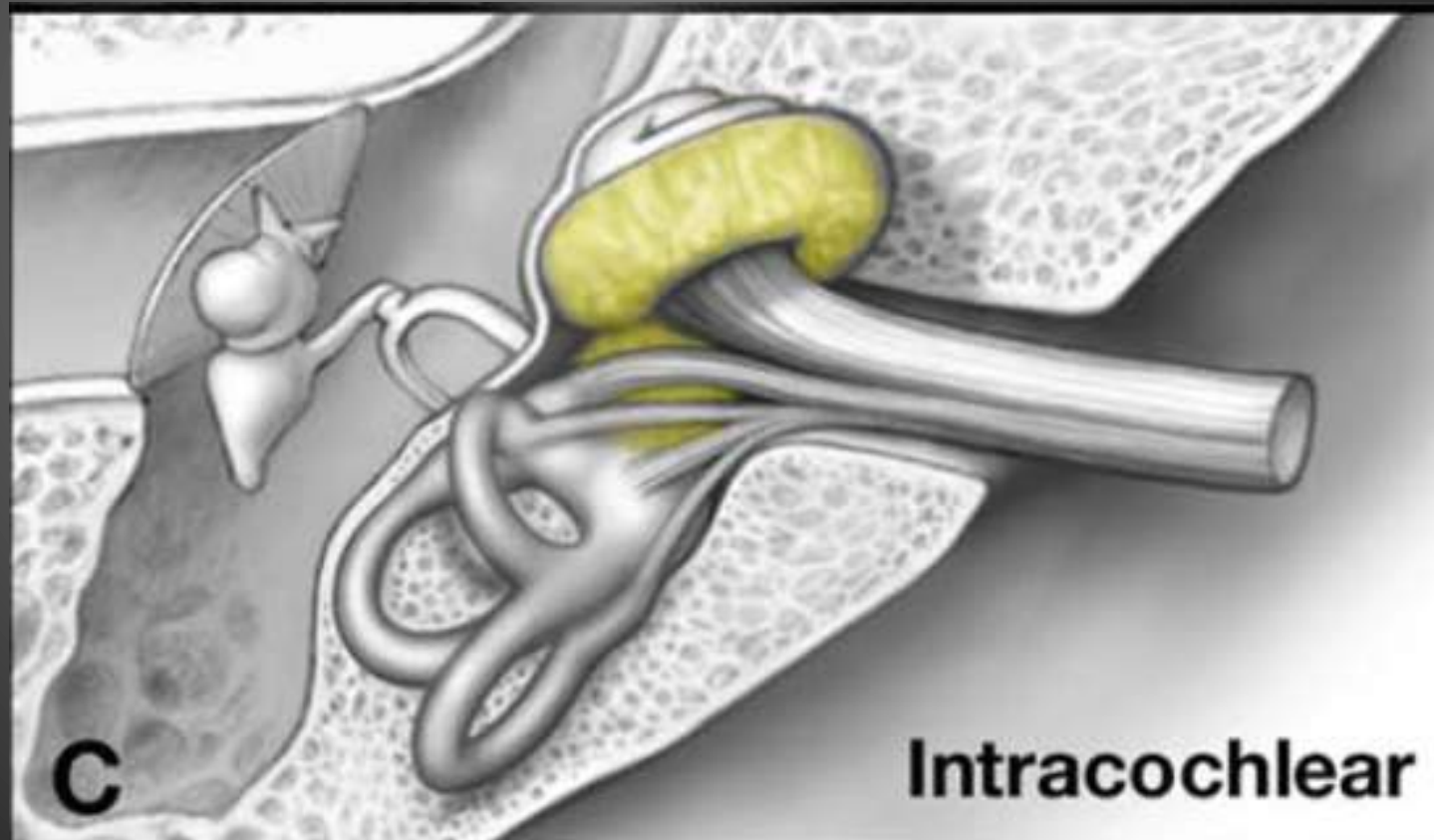
The Kennedy classification includes **intracochlear**, **intravestibular**, **intravestibulocochlear**, **transmodiolar**, **transmacular**, **tympano-labyrinthine**, and **translabyrinthine** tumors

**Table I.** Classification of the Intra-Labyrinthine Chwannomas

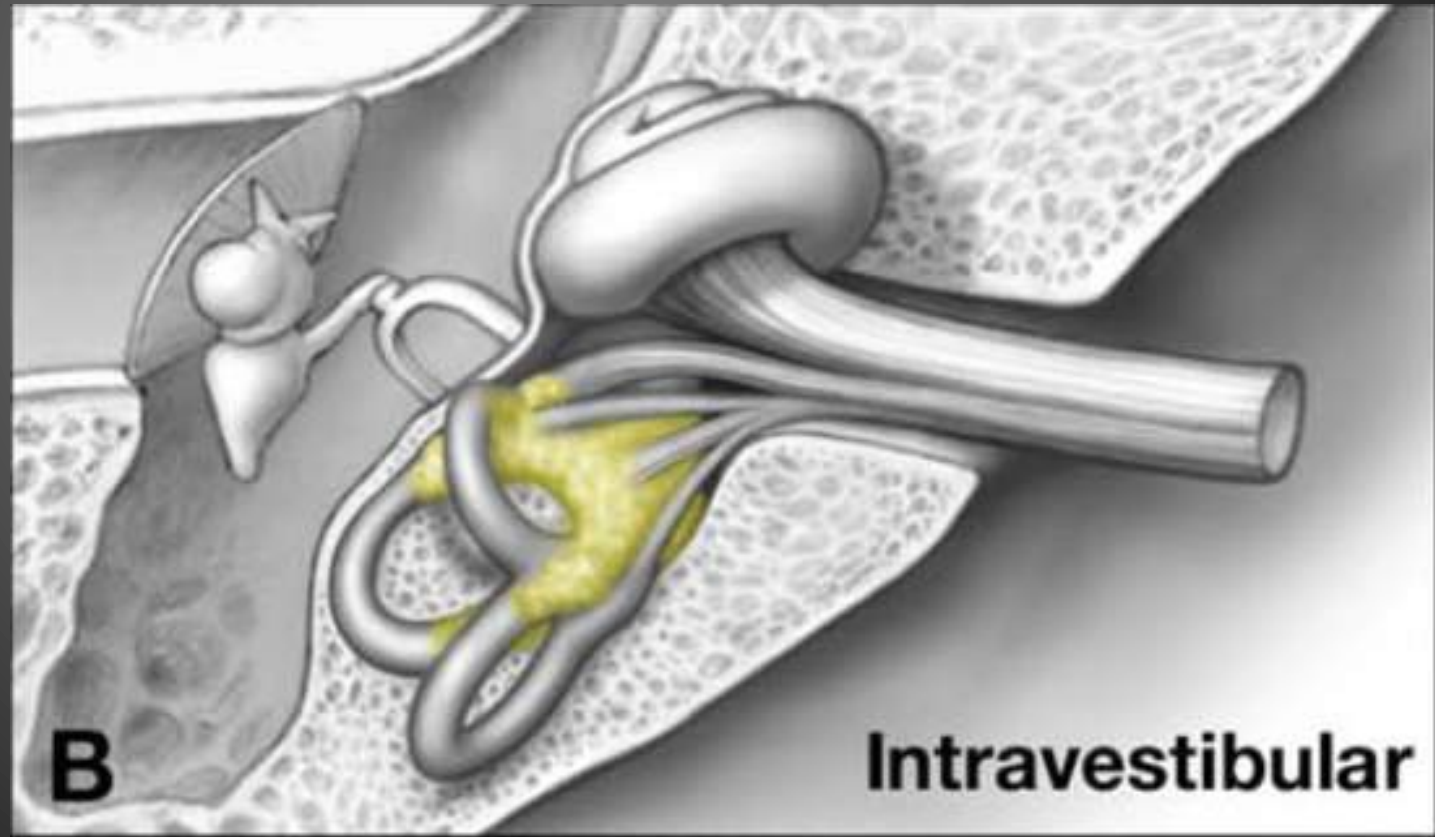
<i>Classification</i>	<i>Area Affected</i>
Intravestibular	Vestibule ± semicircular canals
Intracochlear	Cochlea
Intravestibulocochlear	Vestibule and cochlea
Transmodiolar	Cochlea and IAC
Transmacular	Vestibule and IAC
Transotic	Middle ear, vestibule/ cochlea and IAC
Tympano-labyrinthine	Middle ear and vestibule/cochlea

IAC: internal auditory conduct. Taken from Kennedy et al.<sup>1</sup>

The Kennedy classification includes **intracochlear**, intravestibular, intravestibulocochlear, transmodiolar, transmacular, tympano-labyrinthine, and translabyrinthine tumors

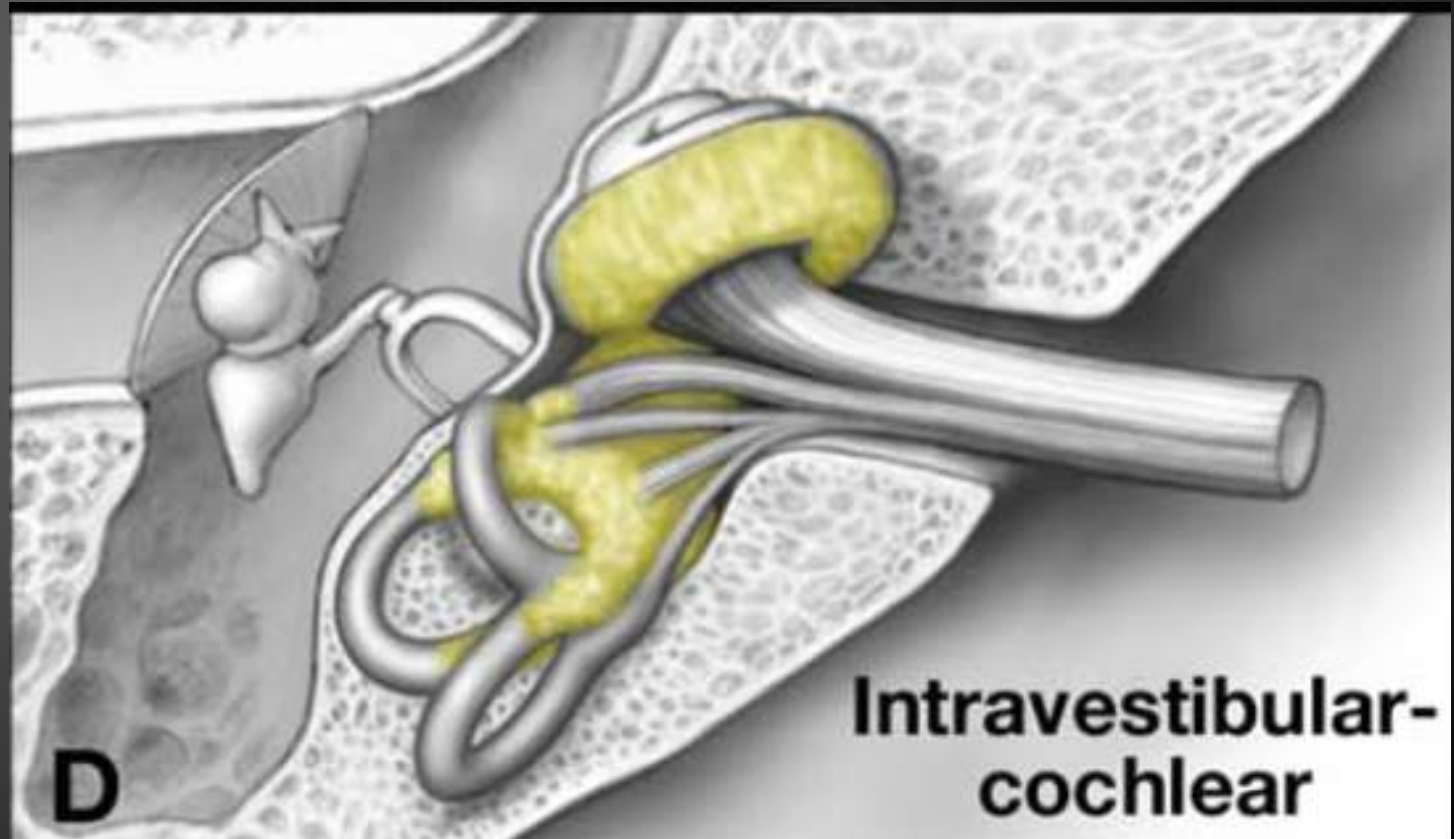


The Kennedy classification includes intracochlear, **intravestibular**, intravestibulocochlear, transmodiolar, transmacular, tympano-labyrinthine, and translabyrinthine tumors

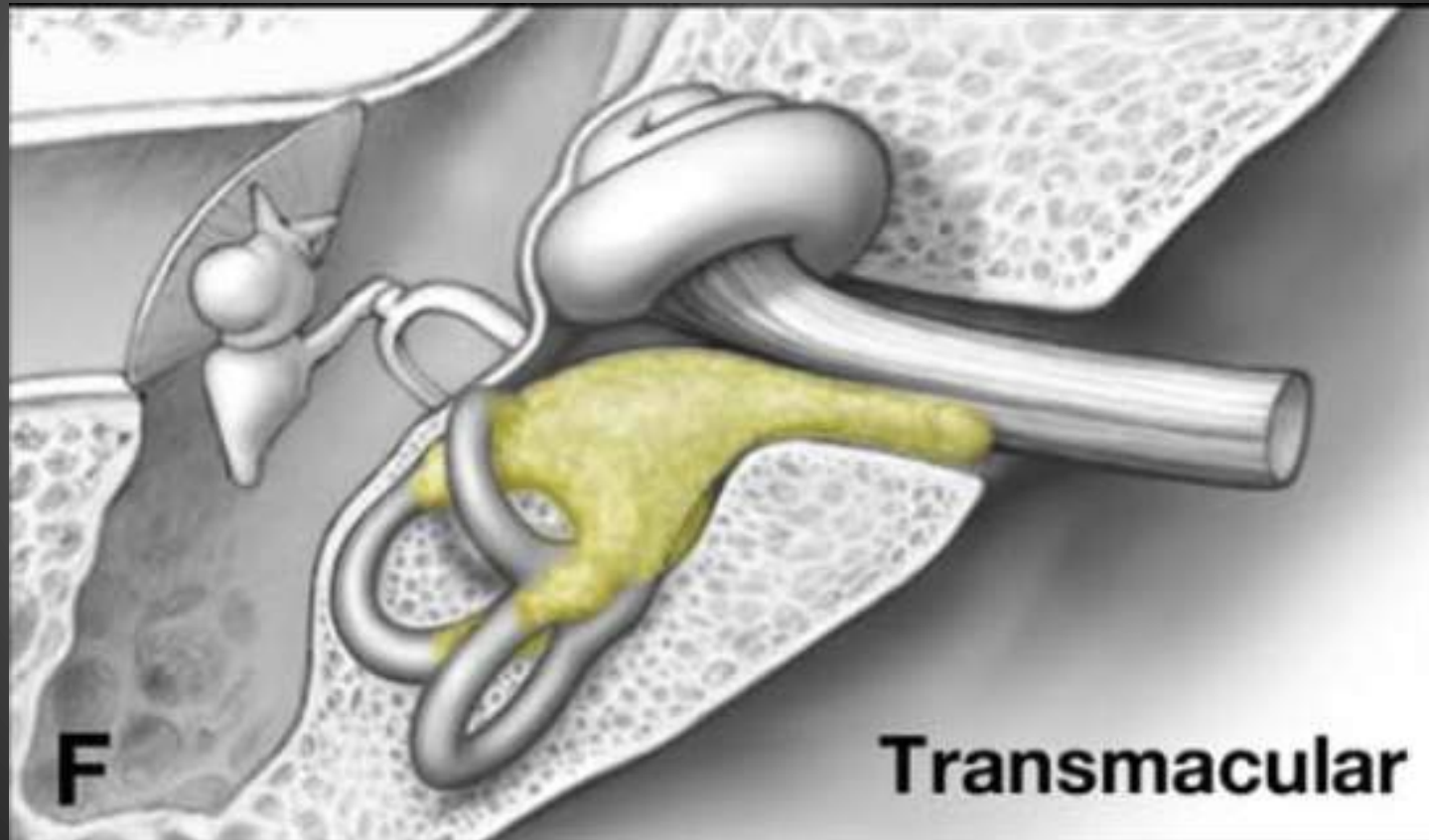




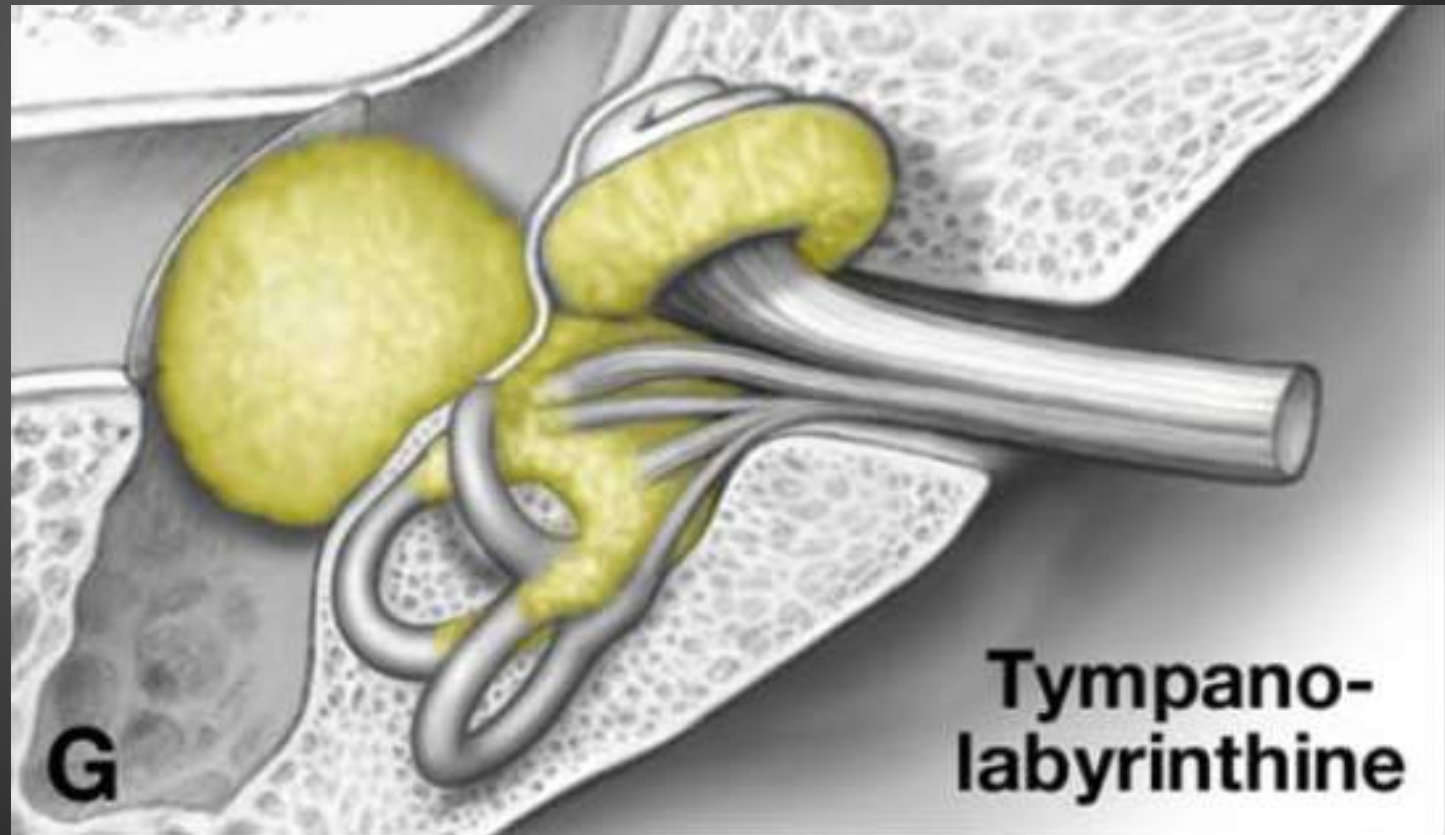
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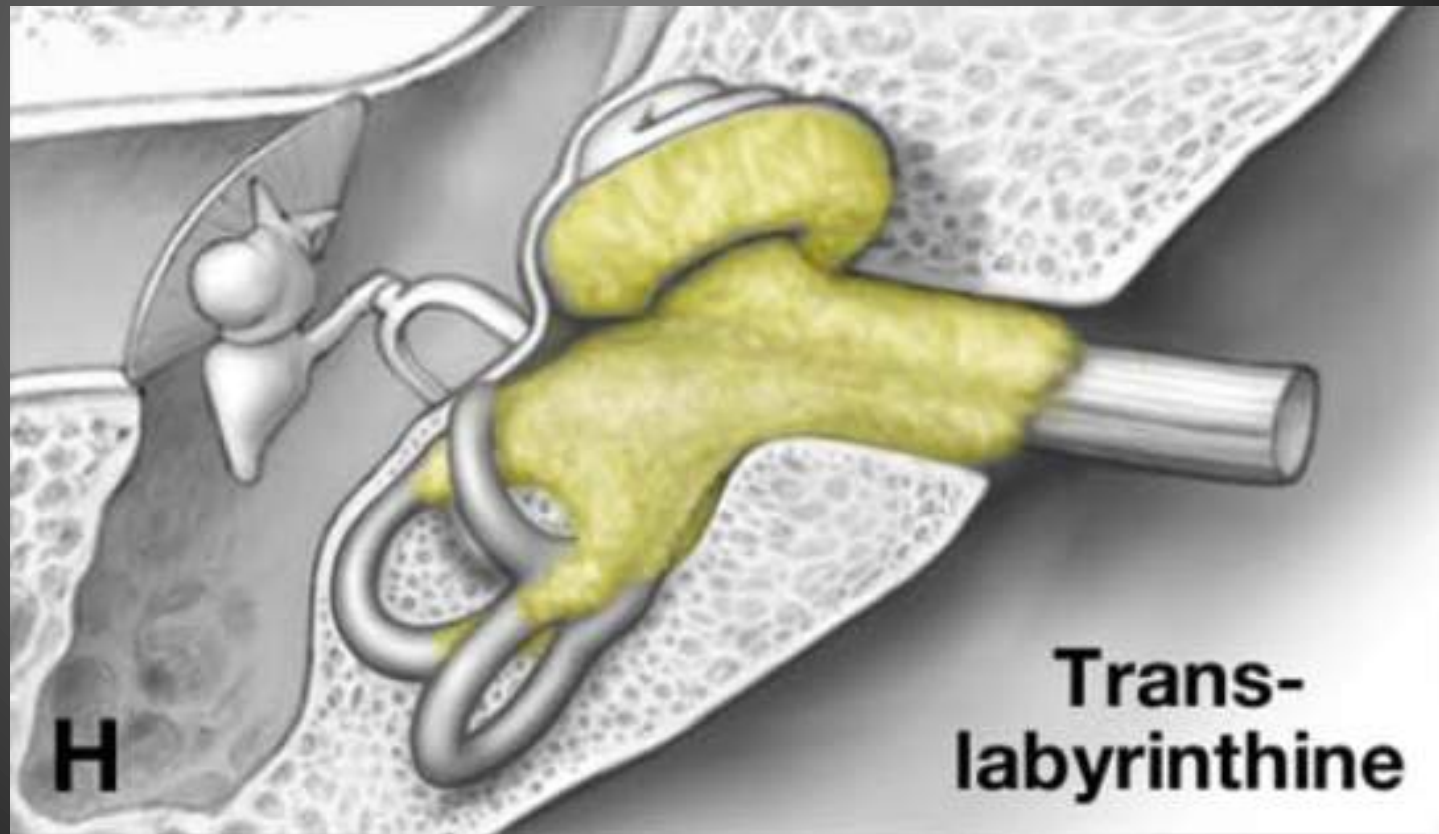
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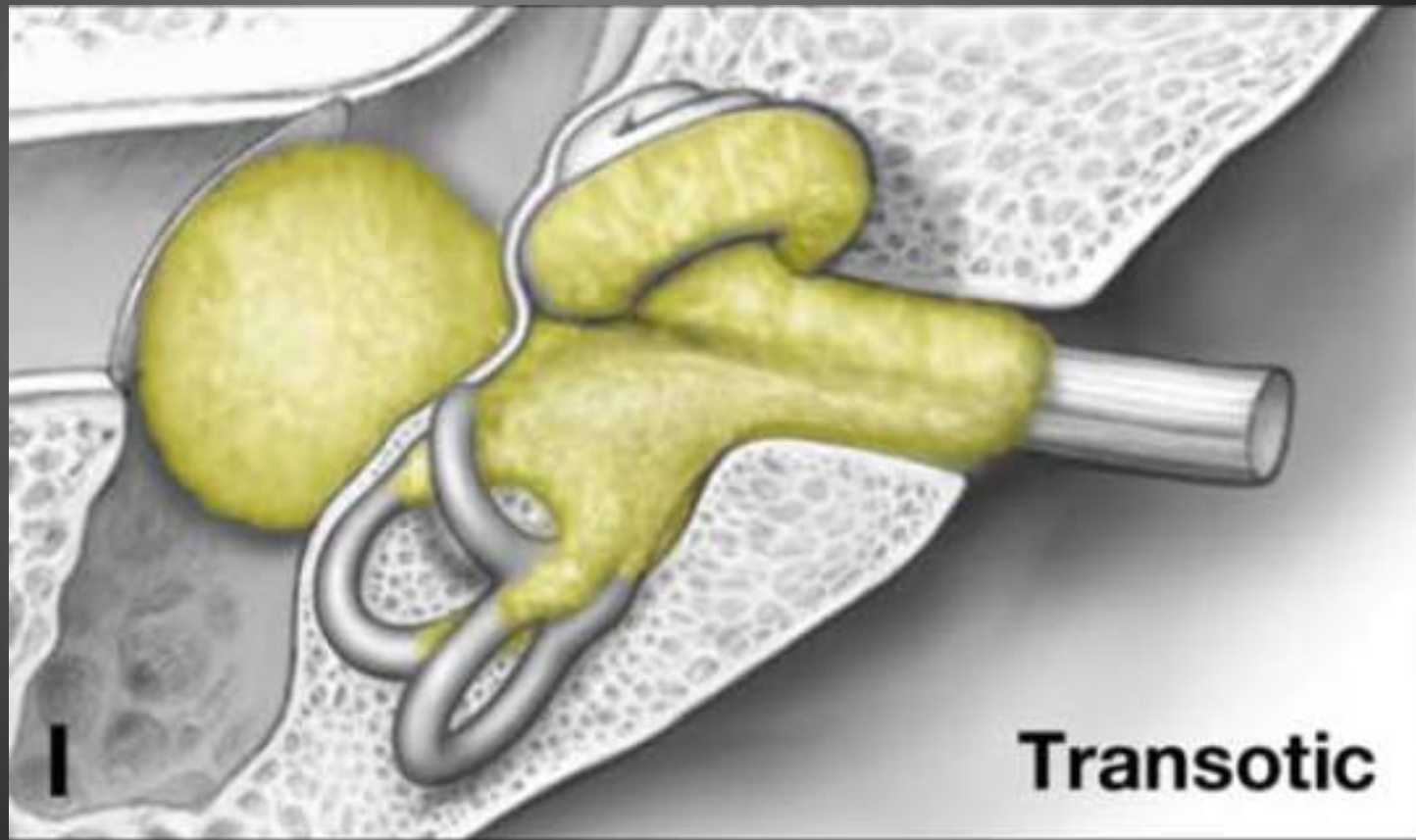
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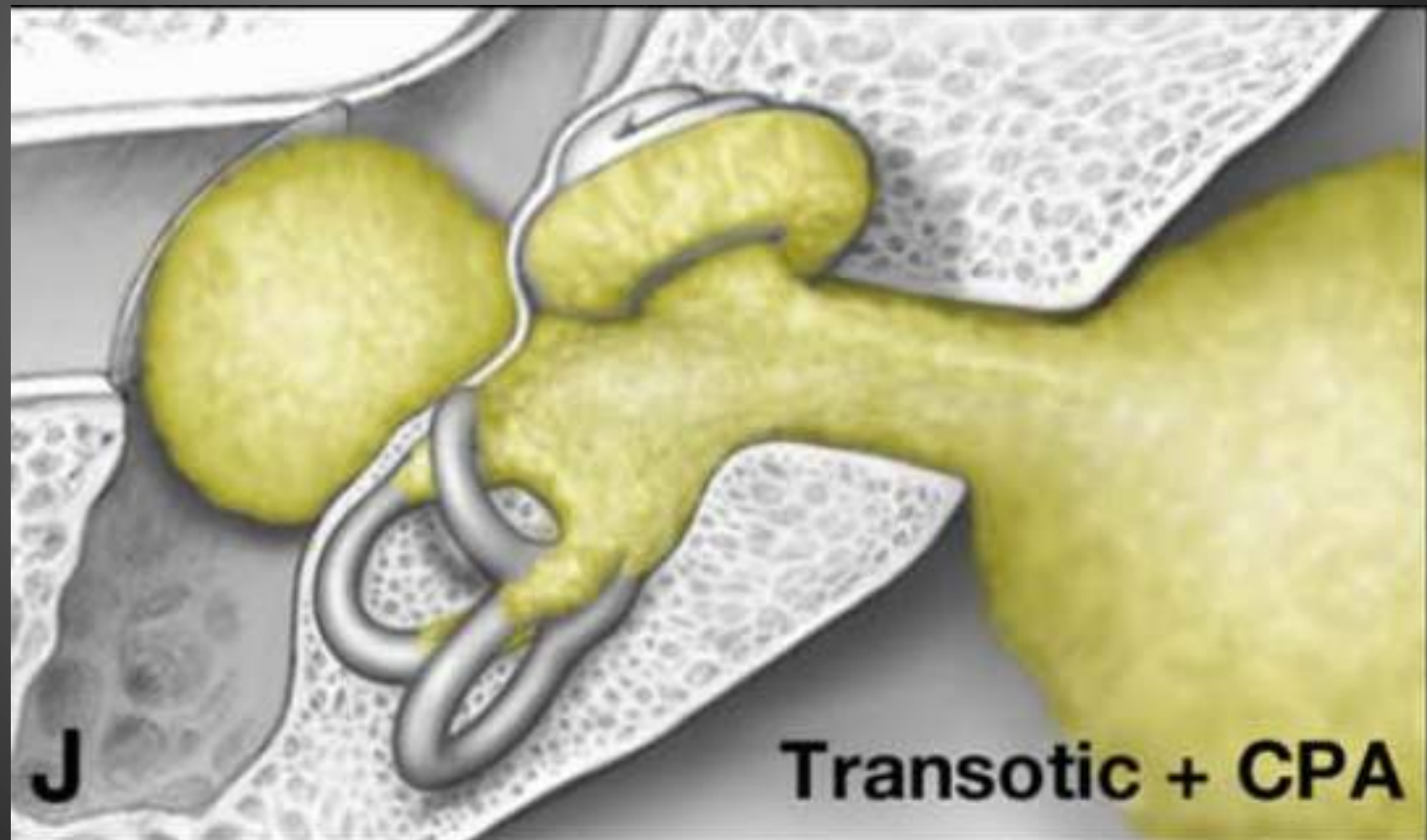
The Kennedy classification includes intracochlear, intravestibular, intravestibulocochlear, transmodiolar, transmacular, tympano-labyrinthine, and **translabyrinthine** tumors



The Kennedy classification system was recently modified also including tumors extending into the cerebellopontine angle



## Modified Kennedy classification



The clinical presentation often includes unilateral hearing loss, vertigo, and tinnitus, mimicking other more common neurotologic conditions.

Second, although high-resolution magnetic resonance imaging has essentially made **early diagnosis** possible in nearly all cases, **a high index of suspicion** is required, as the radiographic findings may be very subtle and easily overlooked.

The symptoms associated with **intra-labyrinthine schwannomas** overlap significantly with other more common neurotologic conditions, such as Menière's disease, vestibular migraine, viral labyrinthitis, and classic vestibular schwannoma, resulting in frequent misdiagnoses.



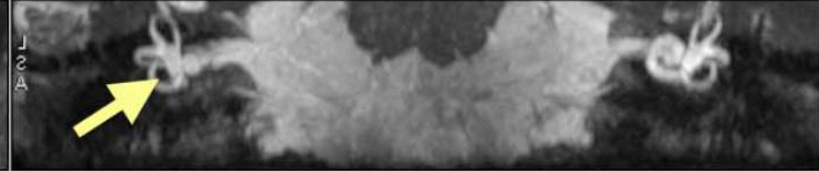
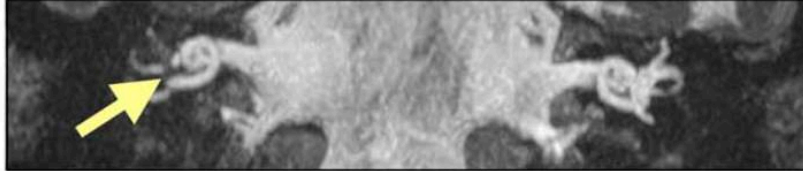
## Intravestibular Schwannoma

## Intracochlear Schwannoma

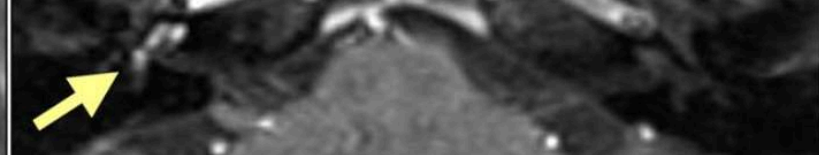
Coronal T1  
+ Contrast



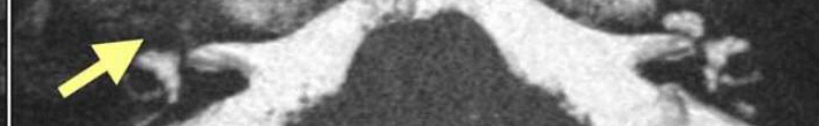
Coronal T2



Axial T1 +  
Contrast



Axial T2



## Clinical Presentation

Unilateral **Hearing Loss** has consistently been the most commonly reported symptom in the **intrala-byrinthine schwannomas** literature.

In the last review, the Authors found that 99% (217/220) of patients presented with some definable **Hearing Loss**

A small subset of patients may experience a **mixed Hearing Loss**, due to an inner ear conductive loss thought to be caused by interference with the intracochlear perilymphatic fluid wave or dampening of stapes movement.

The pattern of **Hearing Loss** (sudden, progressive, or fluctuating) is relevant and may be an important factor in differentiating **intralabyrinthine schwannomas** from Menière's disease.

In contrast to the fluctuating deterioration of hearing, which is the hallmark of Menière's disease, only 3% (5/158) of patients with documented **intralabyrinthine schwannomas** reported a fluctuating pattern.

Tumors involving the vestibule or semicircular canals would intuitively be more likely to cause **vertiginous symptoms** than tumors isolated to the cochlea.

In last review, it found that the symptom of vertigo was strongly associated with tumors involving the vestibular system ( $P < .001$ ).

In addition, there was a statistically significant association between abnormal balance and tumor location; nearly 100% of patients with tumors involving the vestibular system had abnormal findings on physical exam.

## Delay to Diagnosis

**Intralabyrinthine schwannomas** are known to demonstrate an indolent growth pattern, and not surprisingly there is often a significant time interval between initial symptom onset and diagnosis.

In the current studies, the mean age at diagnosis was 48.6 ± 16.8 years (median, 49 years; range, 14–89 years) with an average **delay in diagnosis of 7.0 et al. 8.0 years** (median, 5 years; 0–40 years).

## Management and Prognosis

It is well accepted that the management of **intralabyrinthine schwannomas** is dictated by severity of symptoms and tumor location.

In the literature review it found that a “watch-and-scan” policy was used in 53% of patients (109/189).

Tumor progression was reported in 52% (50/96) of subjects followed with serial MRI.

## Management and Prognosis

Of the patients, 93% (55/59) had either **stable symptoms** or improved tinnitus, unsteadiness, or vertigo regardless of management choice at last follow-up, which supports the conservative watch-and-scan approach to managing these tumors.

Management strategies include microsurgical resection, stereotactic radiosurgery (SRS), or chemical labyrinthectomy.

The primary symptom driving intervention is recalcitrant vertigo, as hearing preservation surgery or SRS is not feasible.

The location of PIES are often associated with both clinical presentation and delay to diagnosis.

An under-standing of vertiginous symptoms and patterns of hearing loss can help differentiate PIES from other more common neurotologic conditions.

MRI investigation is imperative for patients with new onset **asymmetric** sensorineural hearing loss, and close attention must be given toward the inner ear on three-dimensional T2- weighted magnetic resonance cisternography and T1- weighted gadolinium enhanced MRI.



The prognosis for these tumors is excellent, with the majority of patients reporting stable symptoms regardless of treatment.

A watch-and-scan approach is the management strategy of choice in the absence of intractable vertigo or tumor growth beyond the confines of the inner ear.

The most frequent symptom is progressive unilateral sensorineural hypoacusia regardless of whether the tumour is lodged in the vestibular or cochlear branch of the eighth pair.

Balance disorders are less common and their frequency in the different series varies from 25% to 68% of cases.

Treatment is conservative in most patients, and monitoring can be performed with regular resonance imaging.

Surgery is reserved for tumours that invade the IAC and/or the middle ear or in patients with vertigo that cannot be controlled with conservative treatment.

## Ocular vestibular evoked myogenic potentials and intravestibular intralabyrinthine schwannomas

 [Reprints](#)

August 18, 2018 by Massimo Ralli, MD, PhD; Giuseppe Nola, MD; Massimo Fusconi, MD; Luca Sparvoli, MD; Giovanni Ralli, MD

In our case, a 65-year-old man presented with progressive right-sided sensorineural hearing loss, dizziness, and tinnitus and fullness in his right ear.

Audiovestibular examination and MRI detected an intravestibular ILS on the right.

We found that oVEMPs were absent on the contralateral side, which contributed to the diagnostic process. The detection of oVEMPs can provide detailed information on the functionality of the macula of the utricle and the lateral and superior ampullary nerves, with a precise identification of the affected area.

MRI is the current gold standard for its diagnosis, showing lesions with neat margins, hyperintense in T1 and hypointense in T2.

MRI could be used for preoperative localization and surgical planning, as well as for following-up the eventual growth of tumors .

Surgical removal is reserved to a limited number of cases and mostly depends on age, general condition of the patient, tumor size, location and growth (into the internal auditory canal or middle ear), and the presence of intractable symptoms (vertigo).

surgical ablative treatment would result in total hearing loss in 100% of cases, with some likelihood of facial nerve palsy in 4% of cases, cerebrospinal fluid leakage in 5.4% of cases, and meningitis in 1.8% of cases.

In a recent review regarding ILS, among 53% of patients (109 of 189) observed using serial MRI scans, only 3% required surgical removal, supporting the adoption of the wait and scan approach.

In a case, gentamicin infiltration treatment provided excellent results for vestibular symptoms.



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