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Neurotology for Audiologists: a Primer

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Daniel Zeitler, MD, FACS

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Disclosures

- **Presenter Disclosure**: Financial: Daniel Zeitler is an Otologist/Neurotologist at Virginia Mason Medical Center in Seattle, WA. He is on the the Medical Advisory Board Advanced Bionics and a consultant for Cochlear Americas. He received an honorarium for this presentation. Non-financial: Daniel Zeitler has no relevant non-financial relationships to disclose.
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Learning Outcomes

After this course, participants will be able to:

- Describe ABR patterns and vestibular findings in vestibular schwannomas.
- List the characteristics and prevalence of Meniere's Disease.
- Define Autoimmune Inner Ear Disease (AIED) and Sudden Sensorineural Hearing Loss (SSNHL).

Vestibular Schwannomas

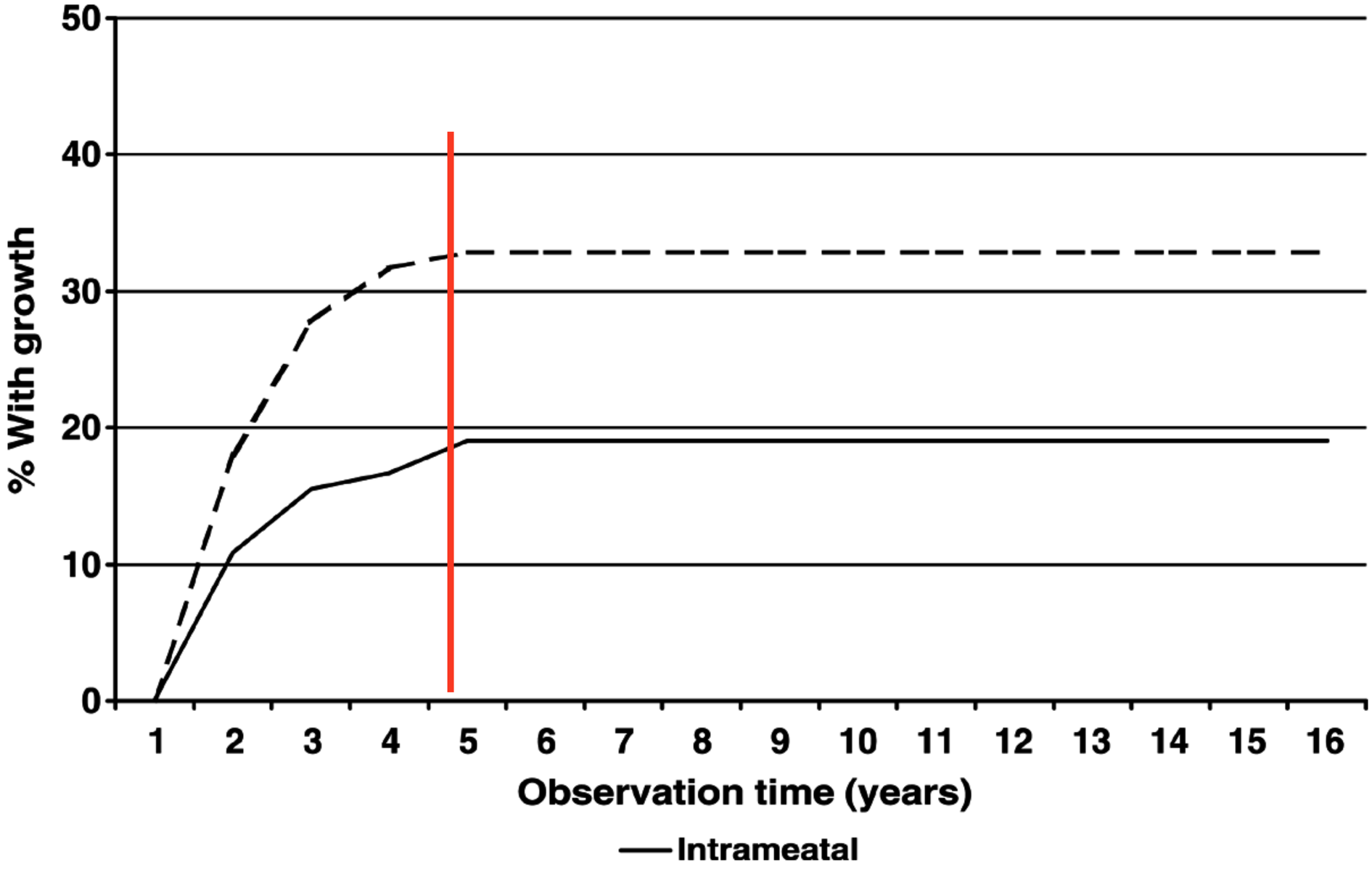
- 6-10% of all intracranial tumors, 80% of all CP angle lesions
- Incidence "1 per 100,000" (0.57%)
- Increasing (MRI technology + access, ASNHL screening)
  - 3-5/100,000, median age 60 yr *(34464224)*
  - Age-specific incidence 20.6/100,000 in ≥ 70 yrs *(32150020; 30032646)*
  - 500/46,400 with symptoms (1%)
  - 8/46,400 (0.017%) incidental VS (mean = 13 mm) *(15781765; 32150020)*
- Lifetime prevalence exceeds 1/500
- 95% sporadic vs. 5% NF2

Vestibular Schwannomas

- Vestibular division of CN VIII
- Equal frequency on SVN and IVN
- Originally thought to arise at Obersteiner-Redlich zone
  - Glial-Schwannian junction
- Linthicum, 2003
  - Scarpa’s ganglion lateral to ORZ
  - Highest concentration of Schwann cells

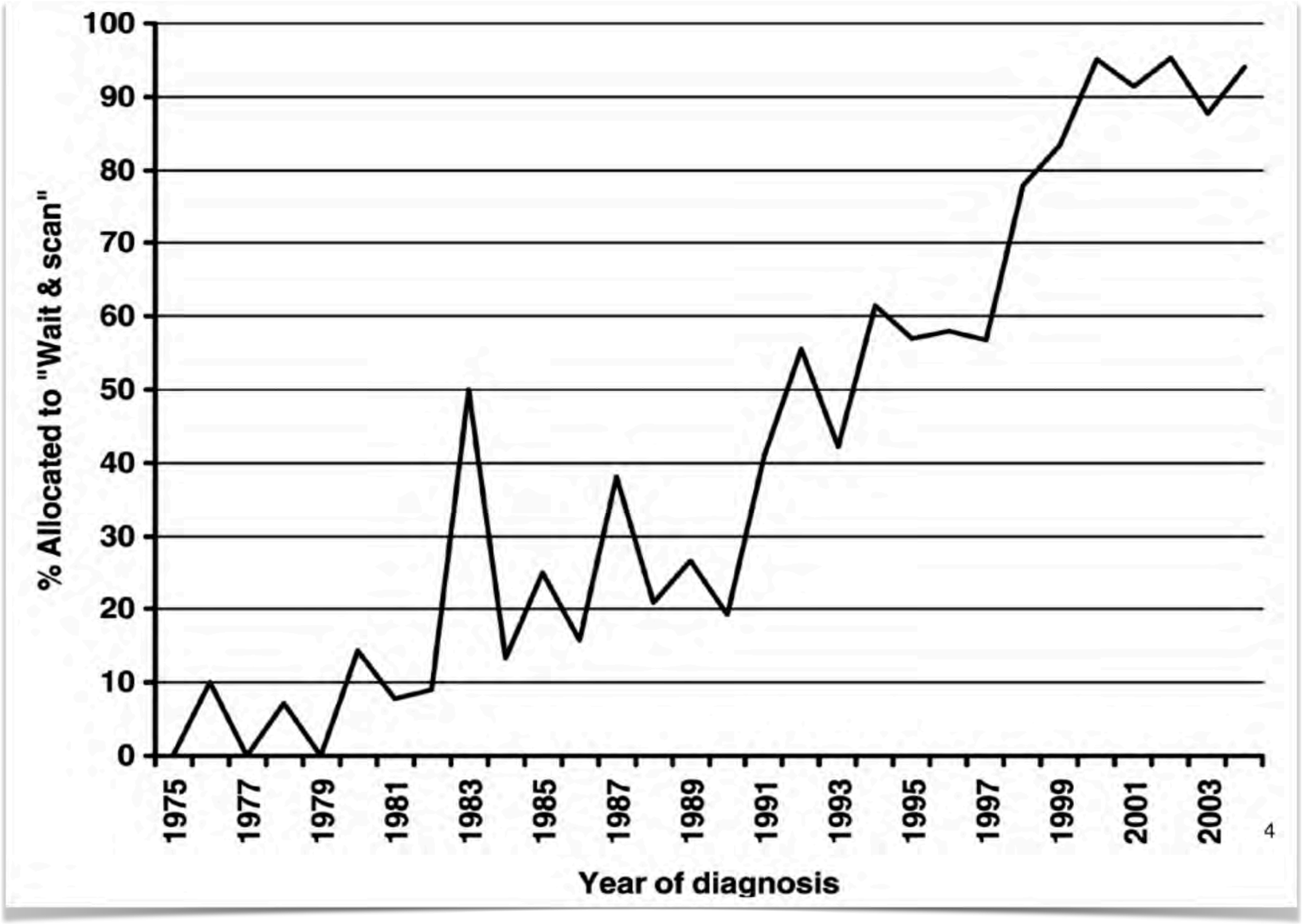
Vestibular Schwannomas: Growth

- Intracanalicular tumors
  - 37% growth rate (10 yr) *(27571523)*
  - Watch and wait failure 15-20%
- EC tumors higher rate of growth (up to 70%) *(27565393)*
- Overall growth 0.6-1.9 mm/yr (82% < 1 mm, 13% > 2 mm) *(16791048)*
- Growth as fast as 3-4 mm/yr in growing tumors


50
40
30
20
10
0
% With growth
1
2
3
4
5
6
7
8
9
10
11
12
13
14
15
16
Observation time (years)
Intrameatal

Results: Follow-up

	Patients with follow-up (n=126)
Mean follow-up time	7.1 yrs (1-16.8)
Mean PTA at last visit (△ PTA)	49.3 dB (11.4 dB)
Mean tumor growth	0.97 x 0.80 mm/yr (25% +growth)
Failed conservative management	14 (11%)



Stangerup SE, *JLO*, 2007

Vestibular Schwannomas: Clinical Manifestations

- Most commonly symptoms isolated to CN VIII (HL, tinnitus, vestibular)
- Cisternal symptoms include facial numbness, hydrocephalus, headaches, imbalance, visual loss
- 85-100% HL, 50-66% tinnitus, 5-60% vertigo, 10-77% CN VII**, 12-29% headache
- Hearing loss often ‘retrocochlear pattern’ - WRS << PTA
- 1% of SSNHL patients with VS, 10% of VS patients with SSNHL

Vestibular Schwannomas: Audiology

Standard audiometry (including rollover and AR decay)

- Rollover - WRS worsens with increasing presentation levels
- Reflex decay – AR decays with prolonged stimulation due to fatigue of auditory nerve
- Neither test sensitive or specific

ABR

- Pattern most specific for VS = + wave 1, no wave 3 or 5; interaural delay
- False positive rate > 80% (low specificity), sensitivity around 80%
- Better for larger tumors (> 1cm)

Vestibular Schwannomas: Hearing

The 4 hearing classes, A to D, of AAO classification		At the last evaluation				
		A	B	C	D	Total
At diagnosis	A	91	53	11	23	178
	B	5	100	83	89	277
	C	—	6	68	89	163
	D	—	2	18	294	314
Total		96	161	180	495	932

1,144 tumors “wait-and-scan”
932 at least 1-year f/u (mean 4.7 yr, 11% 10 year)
51% maintained class A hearing, 54% at 10 years; 13% class D

Stangerup SE, et al, 2010

Vestibular Schwannomas: 100% WRS

The 5 classes, 0–IV, of the mWRS classification		At the last evaluation					
		0	I	II	III	IV	Total
At diagnosis	0	79	59	12	7	2	159
	I	15	137	61	74	45	332
	II	1	15	38	44	29	127
	III	—	7	10	91	71	179
	IV	—	—	3	8	124	135
Total		95	218	124	224	271	932

50% with 100% WRS; 87% better than or = 70%

Stangerup SE, et al, 2010

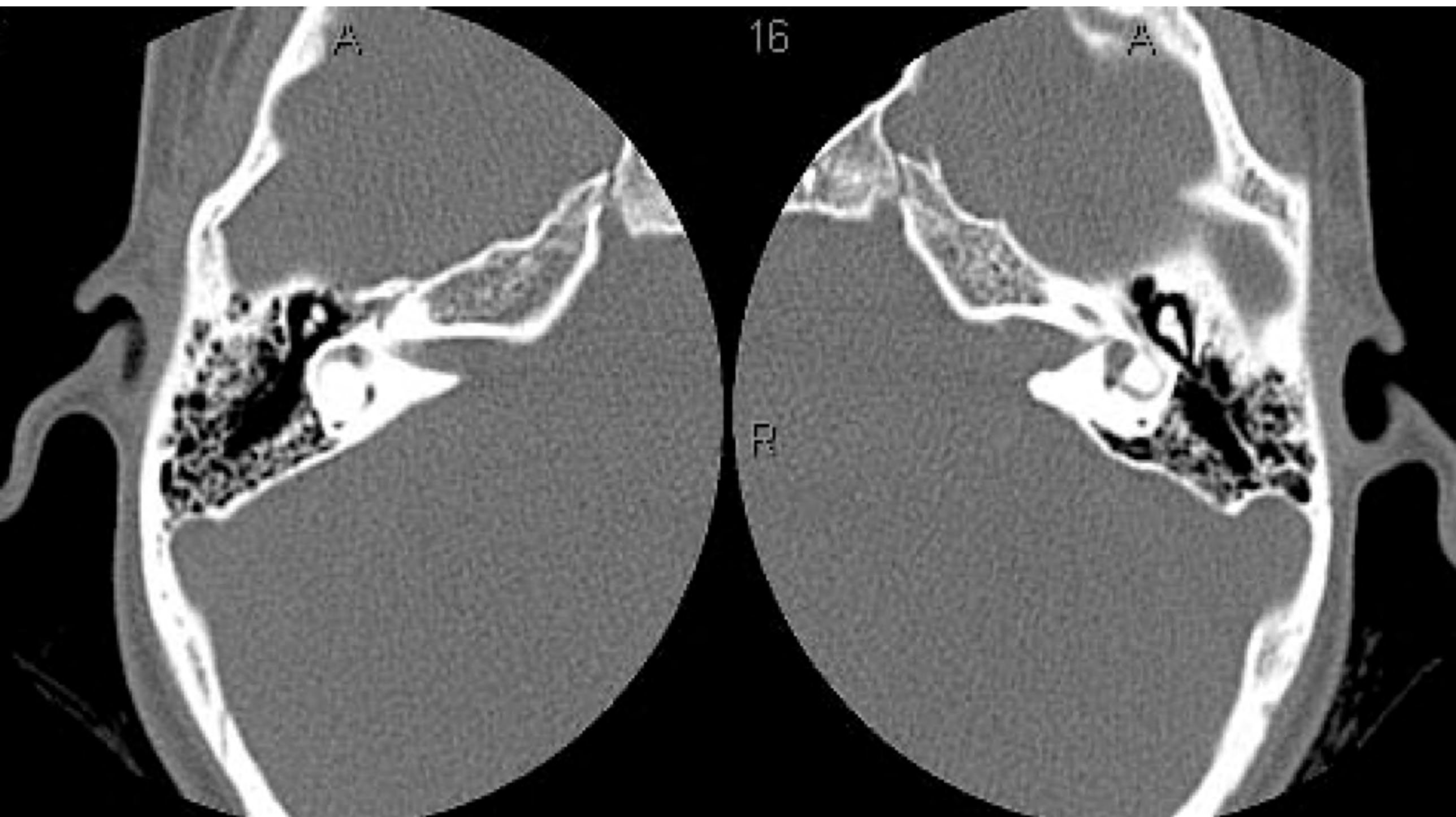
Vestibular Schwannomas: Vestibular Testing

VNG

- Caloric testing = SVN
- SN = 80%, SP = 57%
- 50% absent, smaller tumors often normal response
- Nystagmus with large tumors; beats away from lesion

VEMP

- IVN
- SN = 81%, SP = 57%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A
A
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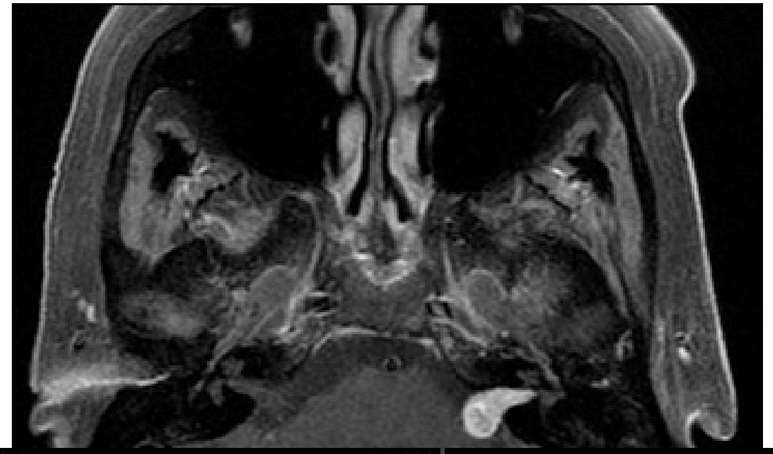

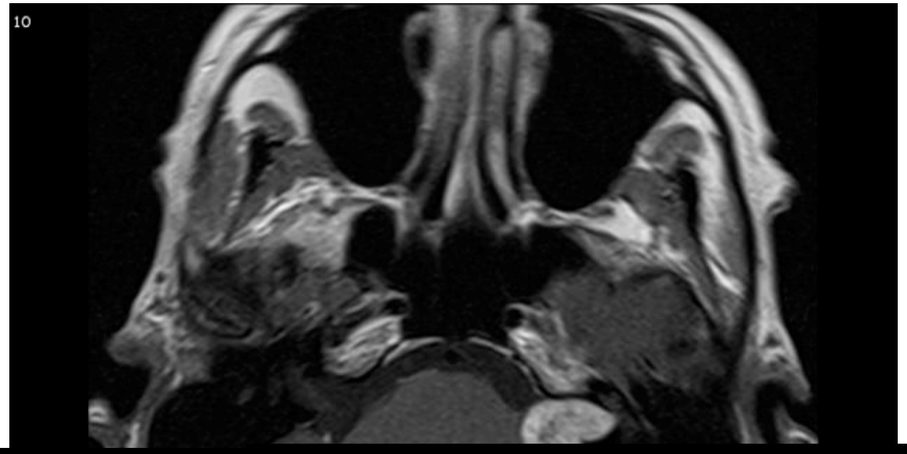
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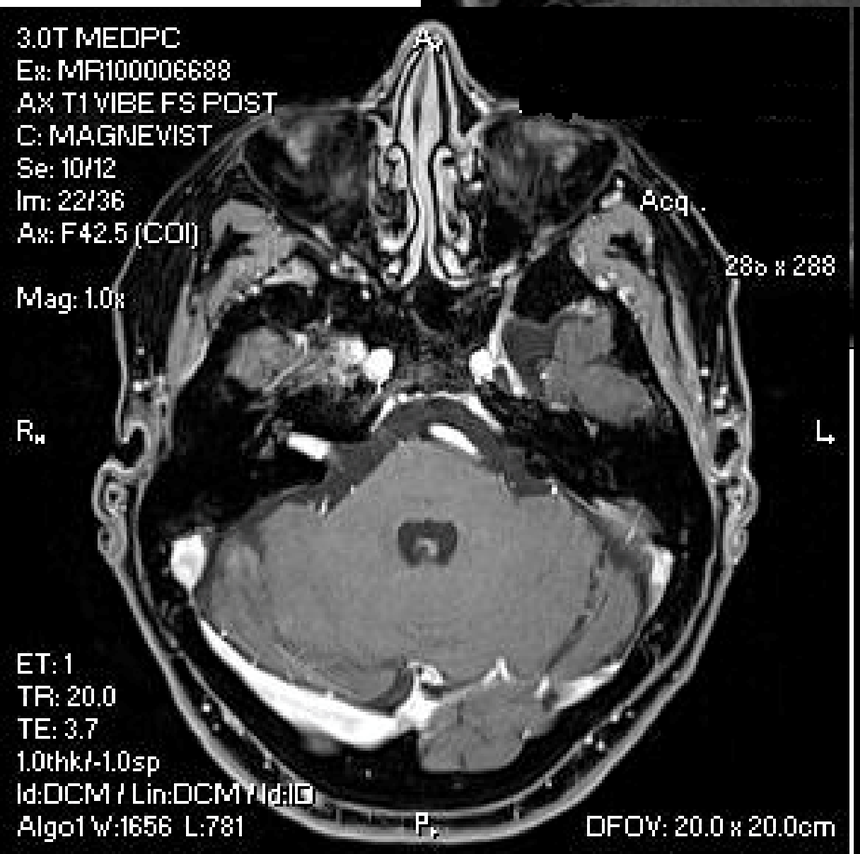
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AX T1 VIBE FS POST
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Im: 22/36
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ET: 1
TR: 20.0
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Algo1 W:1656 L:781
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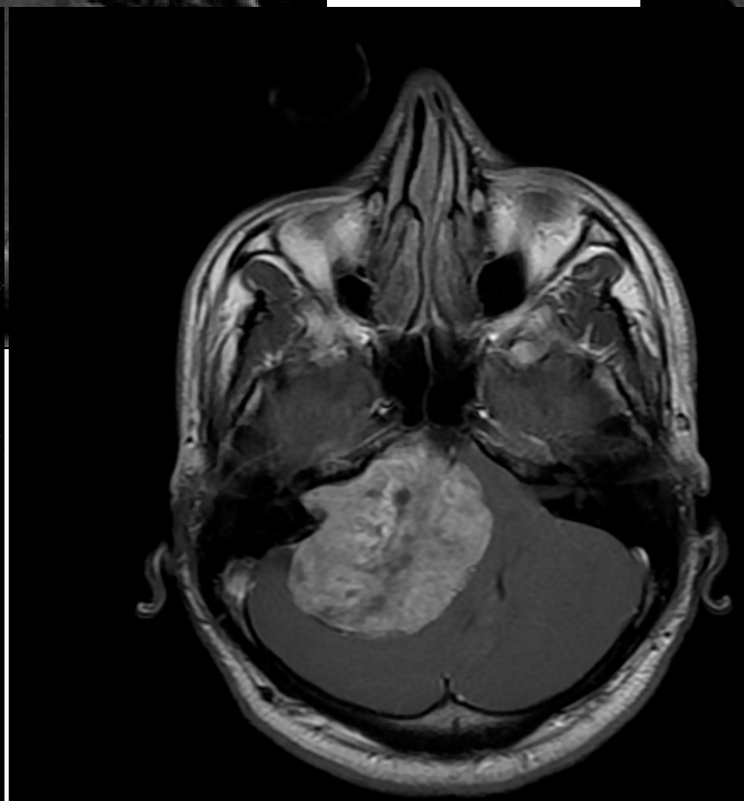
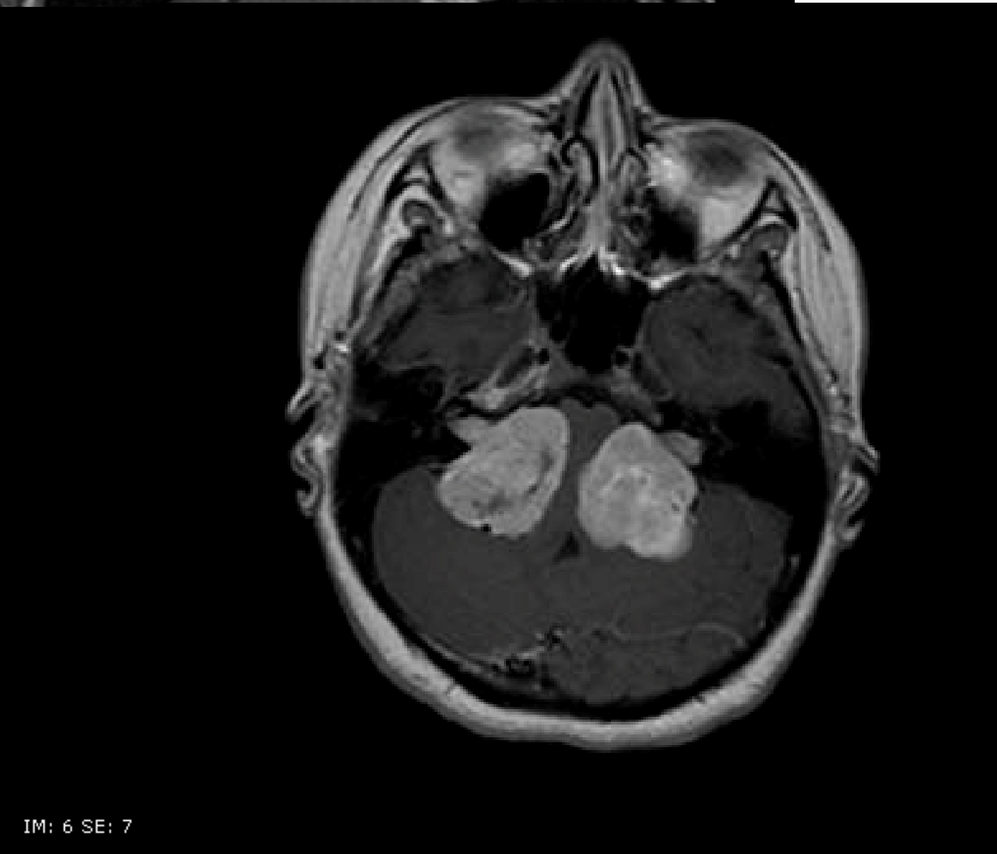
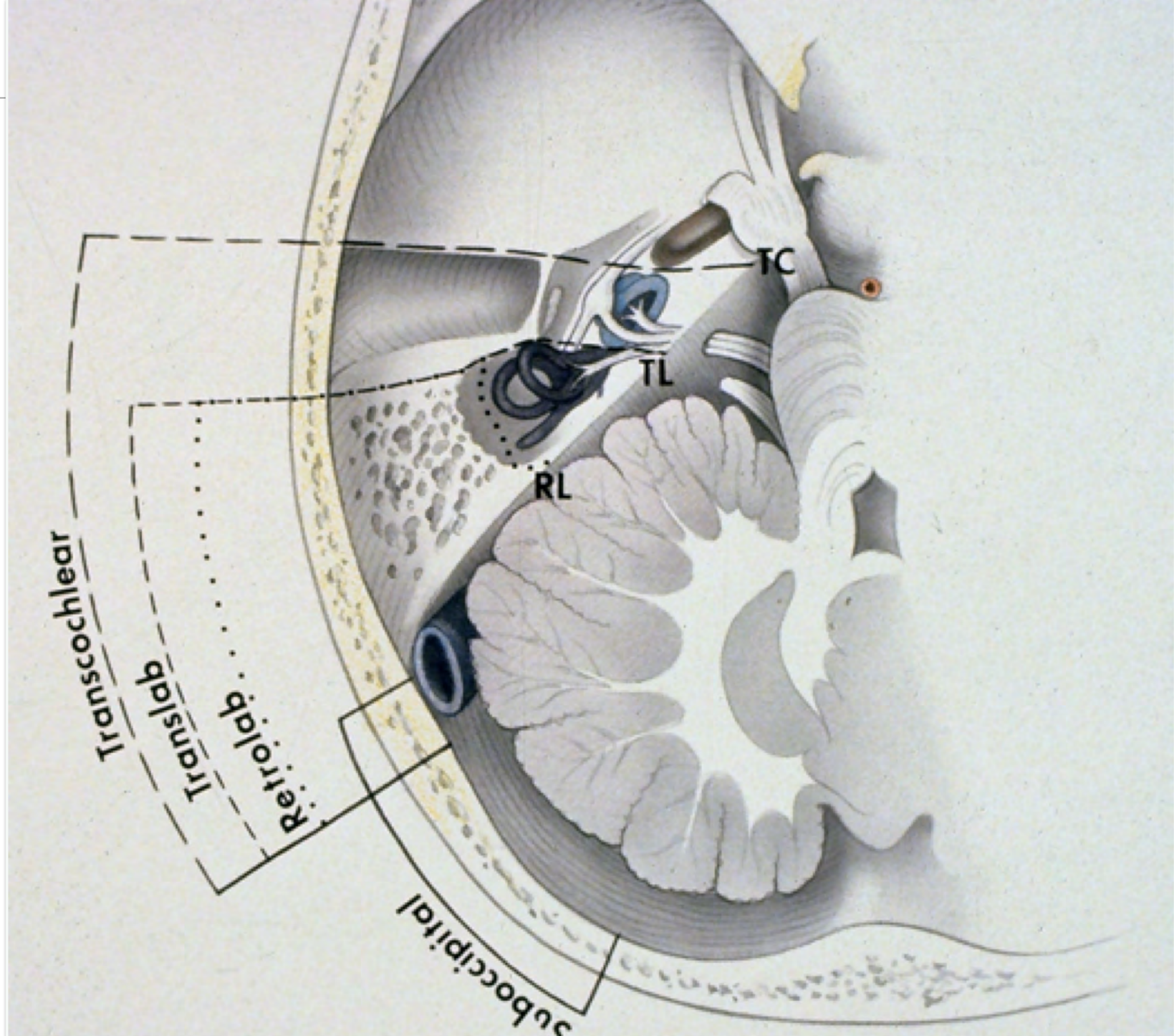




Surgery for vestibular schwannoma



Approach	Indications	Advantages	Disadvantages
Retrosigmoid (Suboccipital)	Any size tumor Any hearing status	Shorter operative time Best wide field view of PCF Good view of inferior CPA Good view anterior to porus acousticus Hearing preservation	May require cerebellar retraction Protracted post-op headache Highest incidence of tumor recurrence or incomplete resection due to poor control of fundus of IAC
Middle Fossa	<1.5 cm tumors Serviceable hearing	Full exposure of lateral third of IAC Hearing preservation	Technically challenging FN may need to be dissected and displaced to obtain access to tumor Limited exposure of PCF Postop trismus due to manipulation and/or injury of temporalis Retraction of temporal lobe Injury of GSPN
Translabyrinthine	>3 cm tumors Smaller tumors with unserviceable hearing	Best view of lateral brain stem Retraction of cerebellum not needed Complete exposure of fundus and lateral end of IAC Better surgical comfort Can identify FN in undistorted form	Hearing is sacrificed Inferior CPA, CNs, and anterior to porus acousticus not well visualized Highest risk of CSF leak Tissue graft required Sigmoid sinus more vulnerable 3rd portion of FN vulnerable to injury

Meniere’s Disease

Meniere's Disease

- Idiopathic and multifactorial
- Accumulation of endolymph in scala media and vestibular organs (endolymphatic hydrops)
- Pathophysiology poorly understood
- Clinical syndrome with episodic vertigo, fluctuating hearing loss, tinnitus, aural fullness
- Disease shows significant variability making phenotyping difficult

Meniere's Disease

- True incidence unknown (subtle onset, fluctuating, long remissions, etc.)
- Estimated 46-100/100,000
- US prevalence 615,000 per NIDCD
- Onset peeks between 40-60 years
- Rare in children
- Bilateral disease 10-12% and increases with time

MD: Old Definition (AAO, 1995)

- **Certain Meniere's Disease**
  - Definite MD + histopathological confirmation (autopsy)
- **Definite Meniere's Disease**
  - 2 or more episodes of vertigo lasting 20+ minutes
  - Documented HL on at least one occasion
  - Tinnitus or aural fullness in affected ear
- **Probable Meniere's Disease**
  - One episode of vertigo
  - Documented HL on at least one occasion
  - Tinnitus or aural fullness in the affected ear
- **Possible Meniere's Disease**
  - Episodic vertigo without documented hearing loss OR
  - SNHL fluctuating or fixed with disequilibrium without definitive episodes of vertigo

International Classification of Vestibular Disorders, 2015 *(25882471)*

Definite Meniere's Disease

- 2 or more episodes of vertigo, 20 minutes - 12 hours
- Low-mid frequency SNHL on at least one occasion, before, during, or after episode of vertigo (at least 30 dB in two contiguous frequencies < 2 kHz)
- Fluctuating aural symptoms in affected ear (hearing, tinnitus, aural fullness)
- Not accounted for by other vestibular diagnosis

Probable Meniere's Disease

- 2 or more episodes of vertigo, 20 minutes - 24 hours
- Fluctuating aural symptoms in affected ear (hearing, tinnitus, aural fullness)

Vestibular Testing: ECochG

- Elevated SP:AP potential due to displacement of basilar membrane toward Scala tympani
- Click stimulus: not reliable; SN 56-67% *(20668393; 15742267)*
- Tone burst: SN 75-85% *(10958398)*
- Poor in early stage disease; better if > 2 years or HL > 30 dB

Vestibular Testing: VEMP

- Presumed saccular dysfunction in MD
- Absent or low amplitude responses in some
- SN < 50%
- SP uncertain 50-95%
- More likely normal in early-stage disease

Treatment: Conservative

- Na+ restriction foundation of medical management
- NO RCTs support this regimen
- Diuretics (HCTZ)
  - Systematic review 4 RCT - 79% found some vertigo reduction *(26932948)*
- Betahistine (Serc) *(23778722; 27697774)*
  - RCT comparing placebo, low dose, high dose - no difference
  - Meta-analysis - **OR 3.37** favoring Serc for vertigo improvement

Treatment: IT therapy

Steroids

- Immune modulation + anti-inflammatory
- Non-ablative, non-ototoxic
- Better vertigo control then placebo

Gentamicin

- Vestibulotoxic, targets dark cells of crista ampullaris in SCC's, utricle, and SV
- Ablative, ototoxic
- 90%+ control of vestibular symptoms

IT Gentamicin vs. IT Steroids

- Casani et al, 2012 *(22101095)*
  - RCT comparing IT Gentamicin vs. IT dexamethasone
  - Substantial control in 92% vs. 61% (complete in 80% vs. 43%)
- Gabra et al, 2013 *(23314159)*
  - RCT comparing IT Gentamicin vs. IT methylprednisolone
  - 83% vs. 48% control rates
- Patel et al, 2016 *(27865535)*
  - IT steroid = IT gentamicin at 18-24 months (90% vs. 87%)

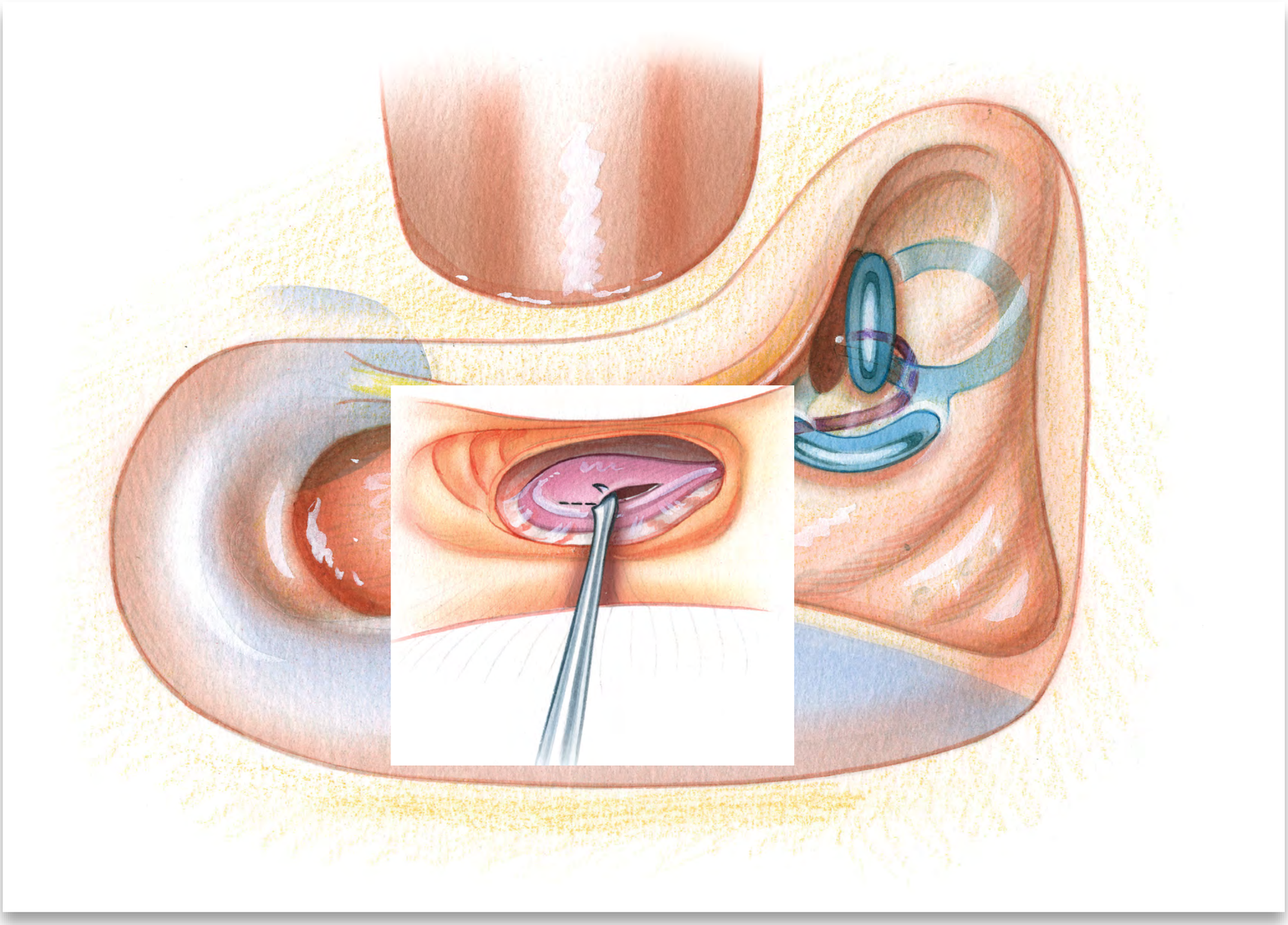
Surgical Options: hearing preservation

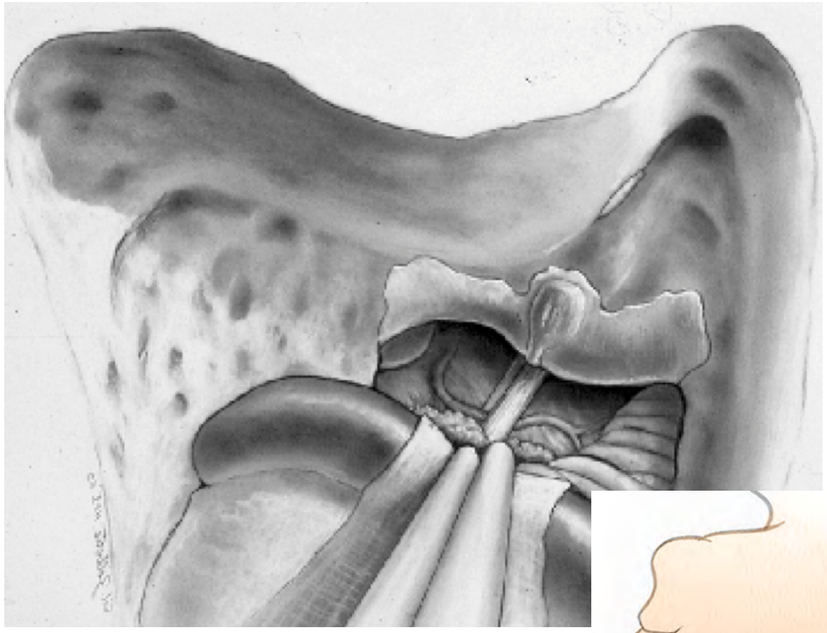
Endolymphatic sac surgery

- Non-ablative, hearing preserving
- RCTs suggest limited efficacy

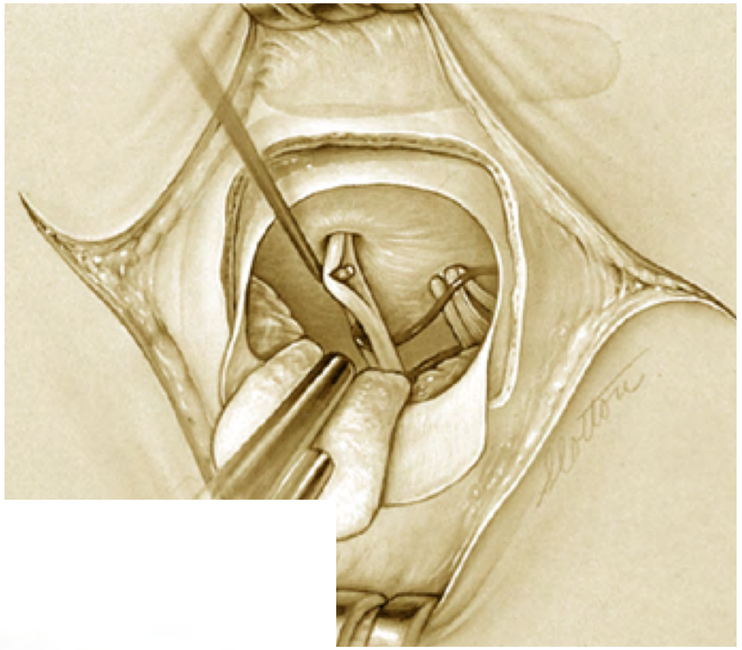
Vestibular nerve section

- Ablative
- Retrolabyrinthine vs. retrosigmoid vs. MCF

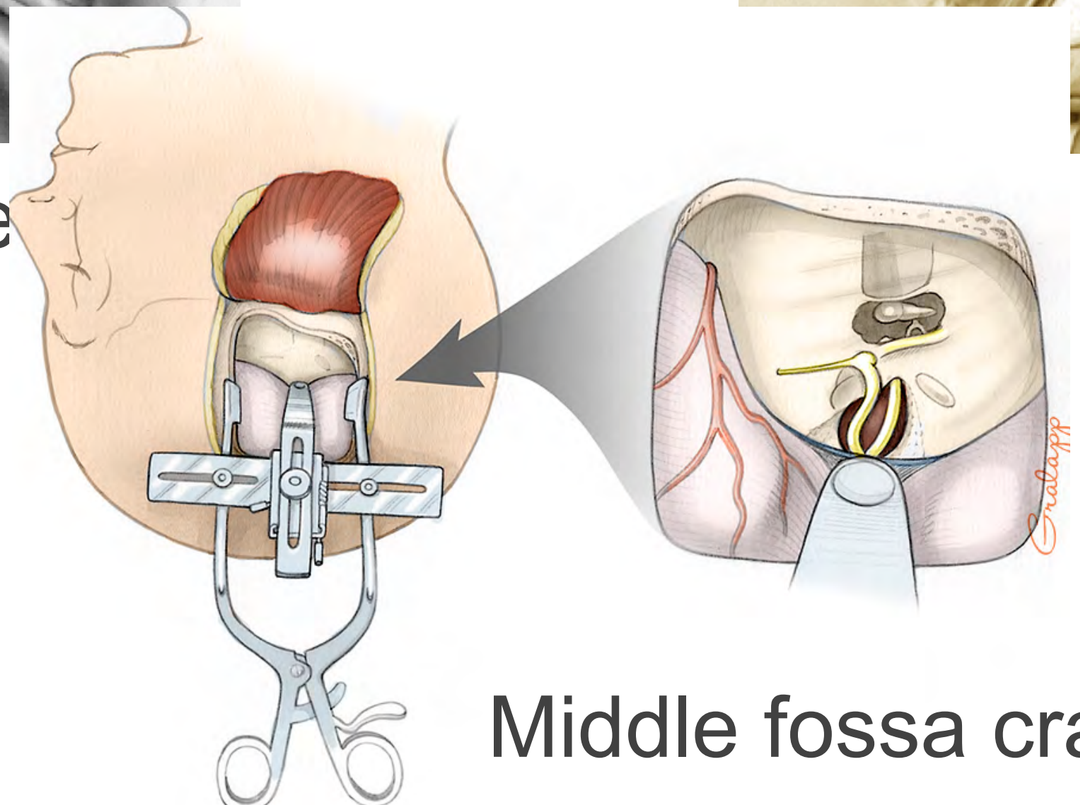




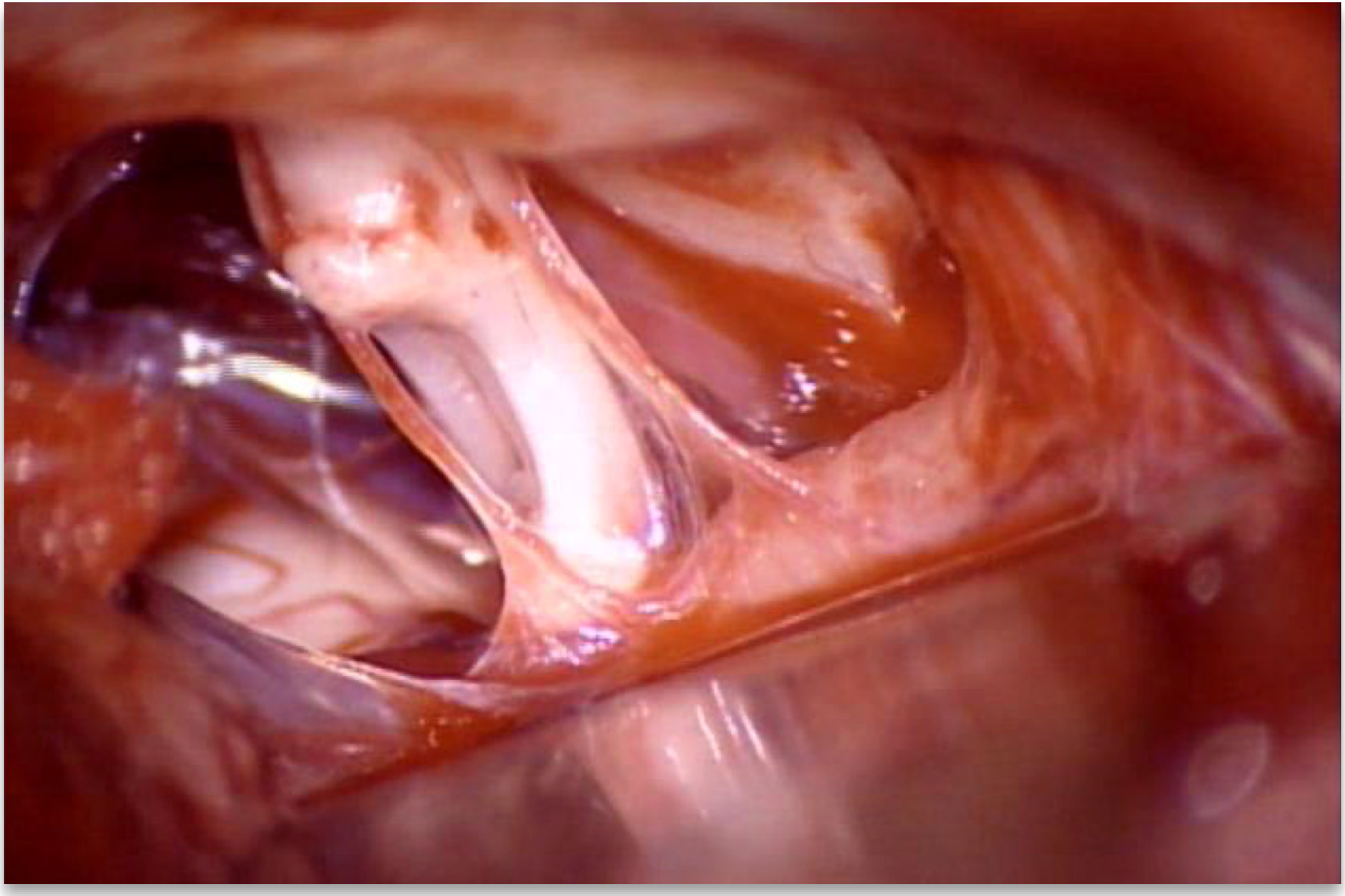
Retrolabyrinthine



Retrosigmoid



Middle fossa craniotomy

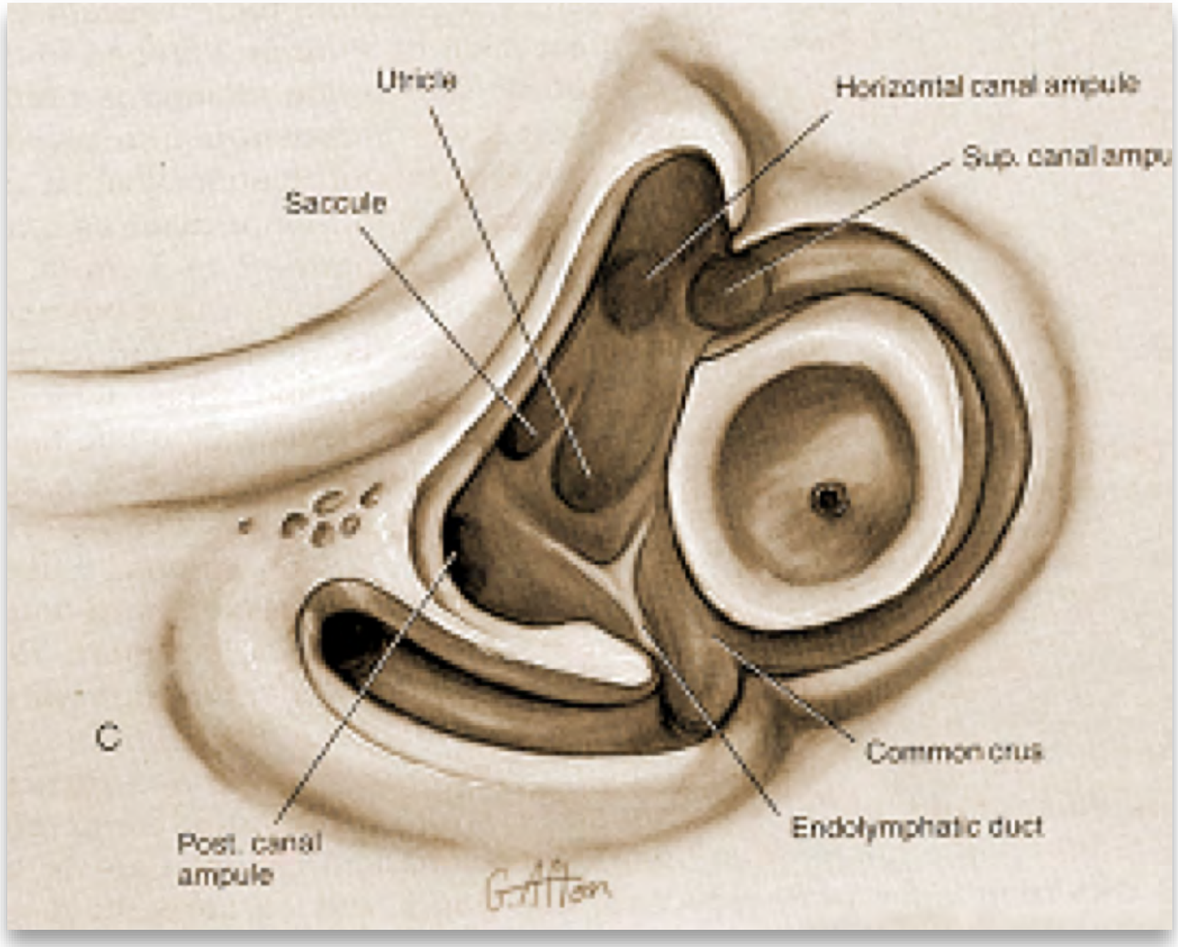


Vestibular Neurectomy Results

Vertigo Control	N	
Silverstein et al 1990	115	94%
Silverstein et al 1990 (review from 58 surgeons)	2590	91%
Li CS, Lai JT 2008	73	94.5%
Hain 2016	(review)	95%

Risks: Hearing loss, risks of intracranial surgery

Labyrinthectomy



		Vertigo Control
Graham (1984)	N=15	93%
Benecke (1986)	N=38	97%
Kemink (1989)	N=110	88%
Langman (1993)	N=26	95%

Autoimmune Inner Ear Disease (AIED)

AIED (IPBSNHL)

- Syndrome of subacute progressive (weeks-months) and often bilateral HL with or without dizziness (50%), tinnitus, aural fullness
- Seen with Lupus, Sjogren's syndrome, Cogan's syndrome, UC, RA, Crohn's Disease, etc.
- Rare (< 1% all cases of HL + dizziness)
- Incidence controversial (16% of bilateral MD?)

AIED: Pathophysiology

- Antibodies or immune cells causing damage to inner ear
  - *Bystander theory*: damage to inner ear invokes immune reaction in cochlea and/or endolymphatic sac
  - *Cross reaction theory*: Ab's or T-cells attack inner ear which shares common antigens with foreign material
  - *Intolerance theory*: antigens released from inner ear may cause immune response
- Genetics?

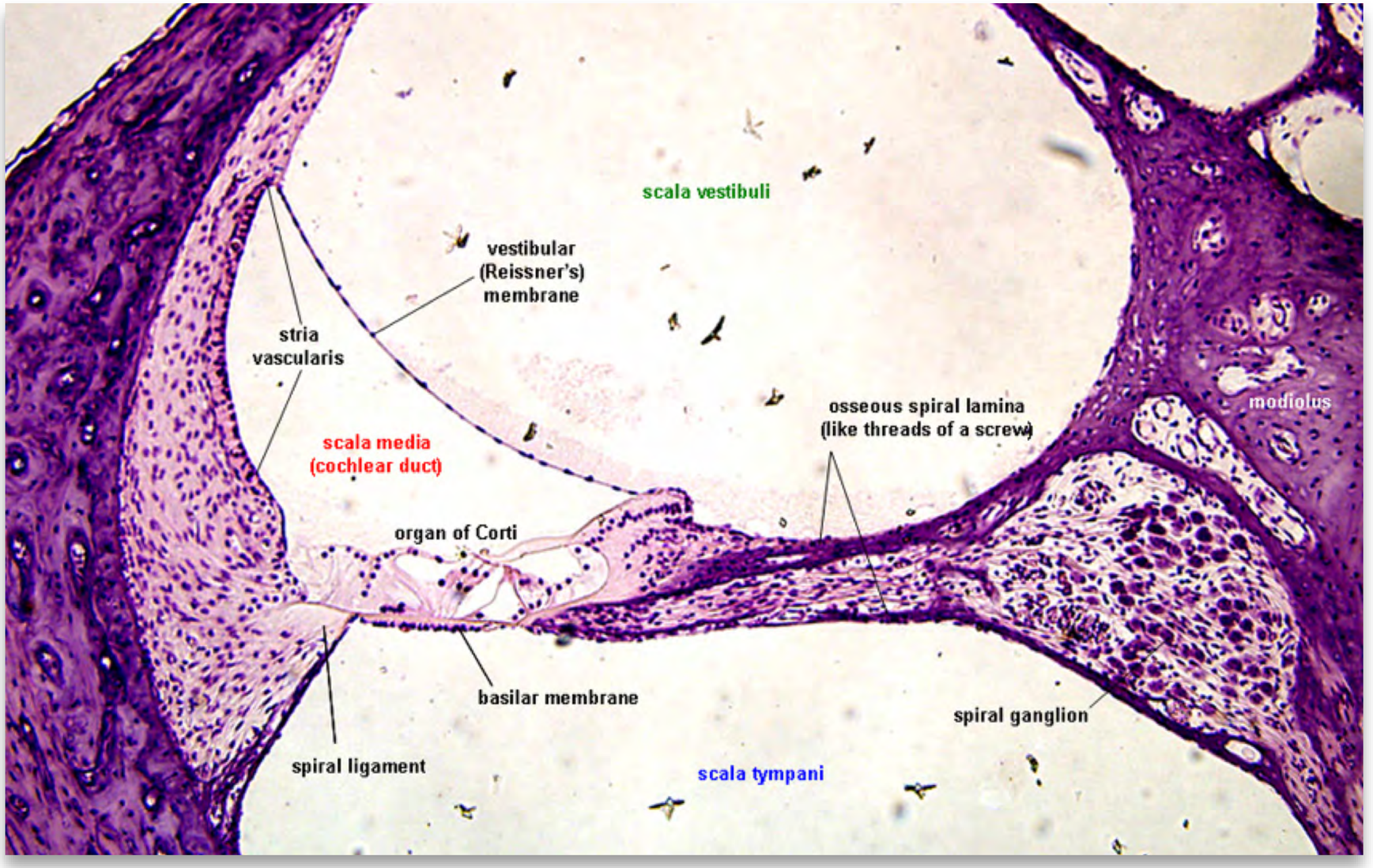
AIED: Diagnosis

- Subacute, classically bilateral SNHL (can be unilateral)
- No characteristic pattern of loss
- **Steroid responsiveness is key to diagnosis**
- Blood tests
  - Anti-cochlear Ab (anti-HSP70) - poor specificity
  - Autoimmune panel (anti-ds DNA, ANA, TPO, RF, anti-gliadin)
- MRI to exclude other causes

AIED: Treatment

- Empiric therapy with prednisone 60 mg QD x 4 weeks
- 60% “responders” (> 15 dB 1 frequency or 10 dB 2 or more, WRS improvement) - transition to non-steroid based treatment after audiogram stabilizes
- MTX classic agent but has been shown ineffective
- anti-TNF (etanercept; Enbrel) (infliximab; Remicade) - $$ *(12544029)*
- Increased risk of lymphoma on anti-TNF agents

Sudden Sensorineural Hearing Loss



SSNHL: Epidemiology

- ≧ 30 dB, 3 contiguous frequencies over 72 hours **(CANNOT BE RIGID)**
- Majority idiopathic - 7-15% with identifiable cause
- Incidence 5-20/100,000 (60,000 cases annually in US)
- 1.5-1.7/100 new patients in otology practice
- Peak incidence 5th-6th decade
- Men = women
- < 2% bilateral, typically sequential

Non-Idiopathic SSNHL

- > 100 possible etiologies
  - Infectious (13%)
  - Otologic (Meniere's)** (5%)
  - Traumatic (4%)
  - Vascular (3%)
  - Neoplastic (2%)
  - Autoimmune**
  - Neurologic
  - Functional

17851927; 8841697; 20422698

SSNHL: Presentation

- Accompanying symptoms:
  - Tinnitus (often roaring) 40-90%
  - Dizziness 30-56%
  - Aural fullness 40-50%
  - Ear popping
- Often noted in the morning upon awakening (vascular hypothesis?)

SSNHL: Natural History

- Proposed etiologies: vascular, viral, intracochlear, immune/inflammatory
- Recovery without treatment = 32-65%
- Spontaneous recovery typically within 2 weeks of onset
- Complete recovery with or without treatment = 36%

Clinical Practice Guideline: Sudden Hearing Loss (Update)

Sujana S. Chandrasekhar, MD[1,2,3], Betty S. Tsai Do, MD[4], Seth R. Schwartz, MD, MPH[5], Laura J. Bontempo, MD, MEd[6], Erynne A. Faucett, MD[7], Sandra A. Finestone, PsyD[8], Deena B. Hollingsworth, MSN, FNP-BC[9] , David M. Kelley, MD[10], Steven T. Kmucha, MD, JD[11], Gul Moonis, MD[12], Gayla L. Poling, PhD, CCC-A[13], J. Kirk Roberts, MD[12], Robert J. Stachler, MD[14], Daniel M. Zeitler, MD[5], Maureen D. Corrigan[15], Lorraine C. Nnacheta, MPH, DrPH[15], and Lisa Satterfield, MS, MPH[15]

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(1) **Strong recommendation**: exclude conductive hearing loss

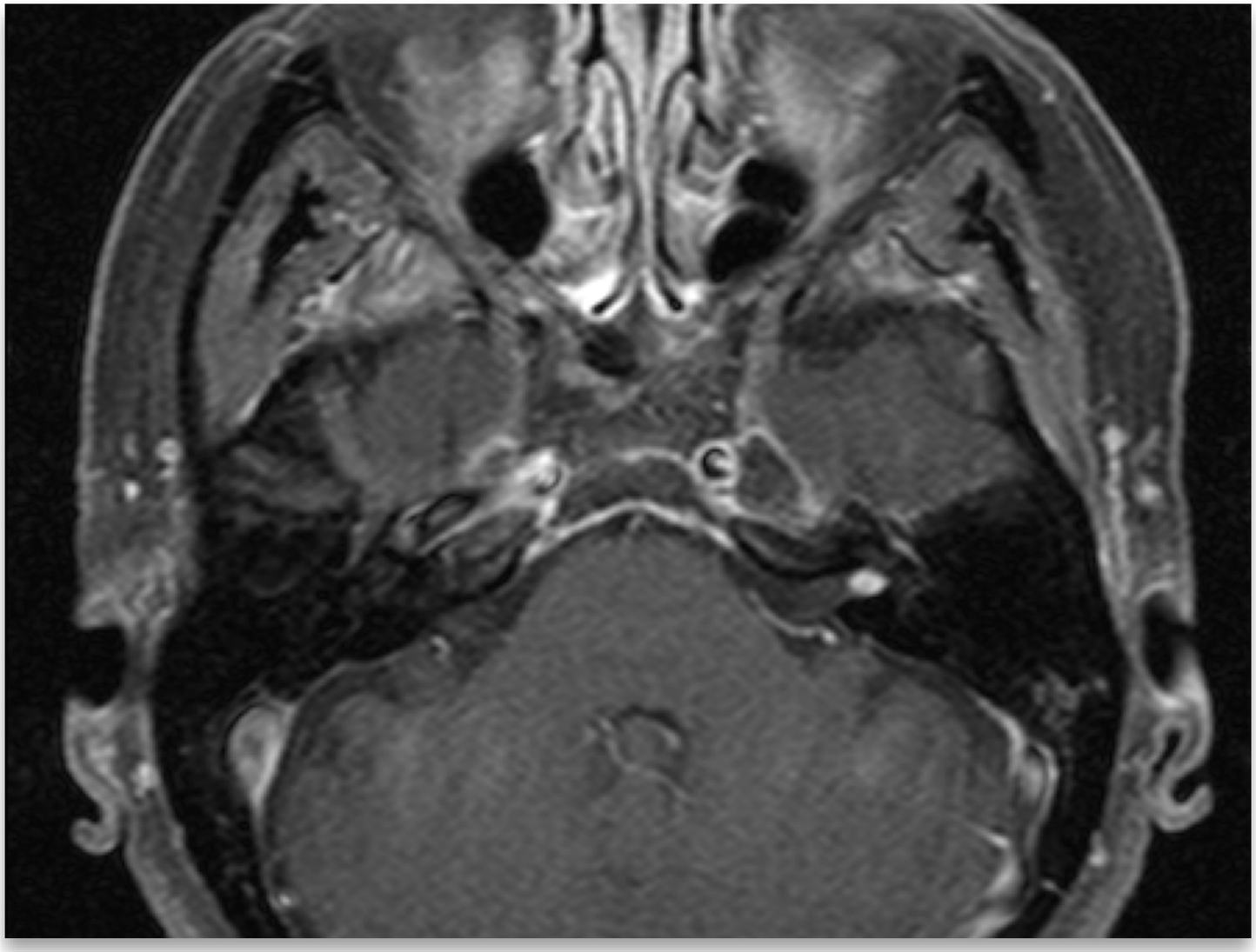
(2) **Recommendation**: assess for modifying factors (bilateral SNHL, neurological findings, recurrence)

(3) **Recommendation**: audiometric confirmation

(4) **Strong recommendation against**: laboratory testing

(5) **Strong recommendation against**: CT scanning

(6) **Recommendation**: MRI (1% VS vs. 1/100,000)



Sensitivity and specificity of MRI for tumors > 2mm nearly 100%

SSNHL: Steroids as Initial Treatment

Option: Steroids

98% US Otolaryngologists treat with steroids *(19130968)*

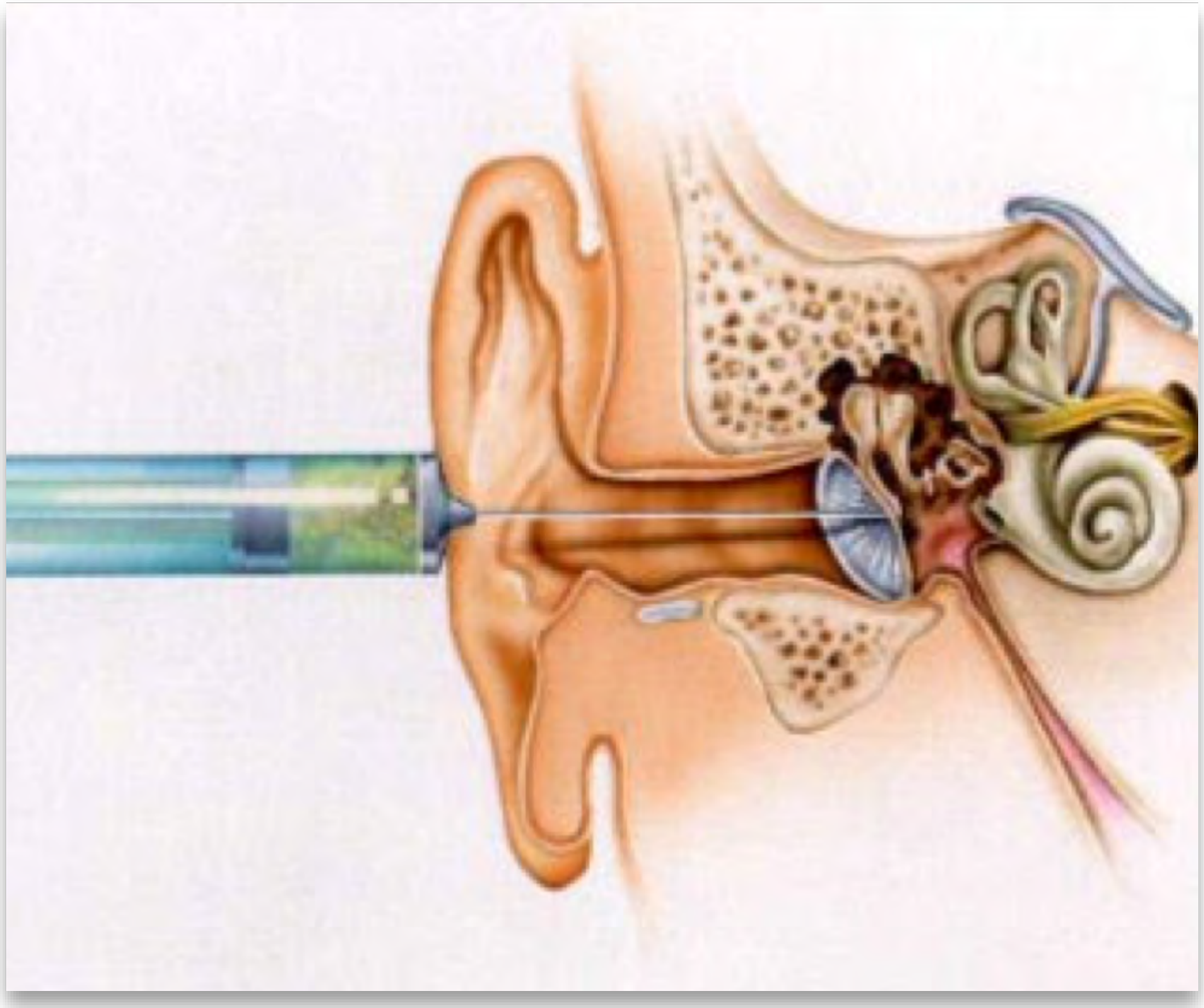
Does the literature support steroids for SSNHL?

- PubMed: 1980-2014: 491
- 3 meta-analysis

	Location	Articles	Conclusion
Conlin & Parnes, 2007	Ontario	2	Unclear benefit of steroid vs. placebo
Labus et al. 2010	Brussels	6	Medical therapy > placebo, not significant
Wei et al. 2013	Melbourne	3	Unclear benefit of steroid vs. placebo

SSNHL: IT Steroids as Initial Treatment

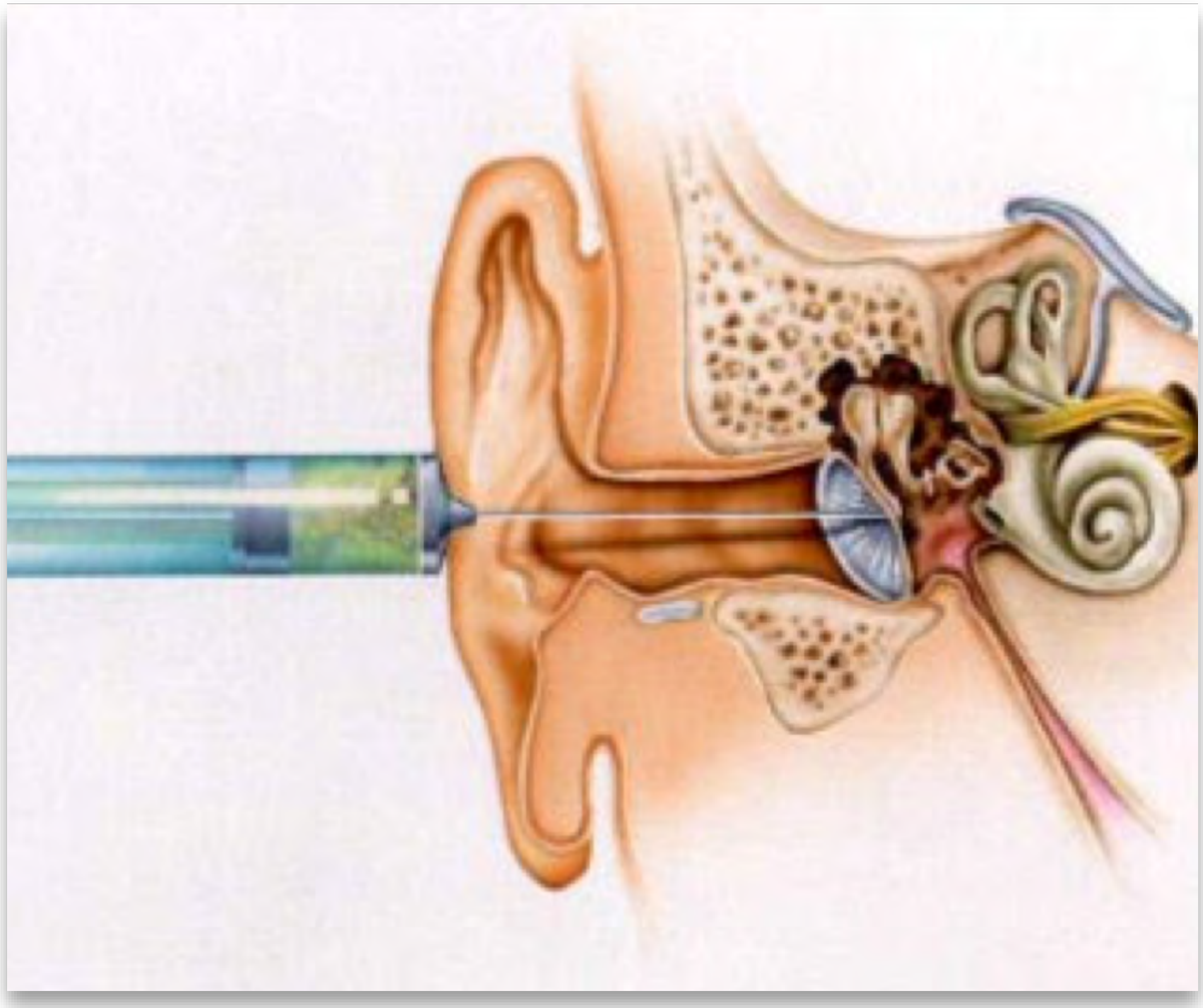
- Intratympanic steroids
  - Little systemic absorption
  - Patients who cannot take oral steroids (DM)
  - Higher intracochlear concentration
  - **Not inferior** to PO steroids as initial treatment *(21610239; 21327728)*



SSNHL: IT Steroids as Salvage Treatment

- 5 RCT - all showed significant benefit after failed systemic therapy
- Dexamethasone 10 mg/mL - 4 shots over two weeks
- OR 6.04 but audiometric "*success*" not defined

RECOMMENDATION



(15235345; 16730534; 22339625; 21646929; 21221620)

SSNHL: Hyperbaric Oxygen

- Increase oxygen levels in cochlea if vascular/ischemic insult
- Greater improvement with less severe initial loss
- Greater improvement if treated under 2 weeks
- NNT =5 for > 25% improvement in PTA *(23076907)*

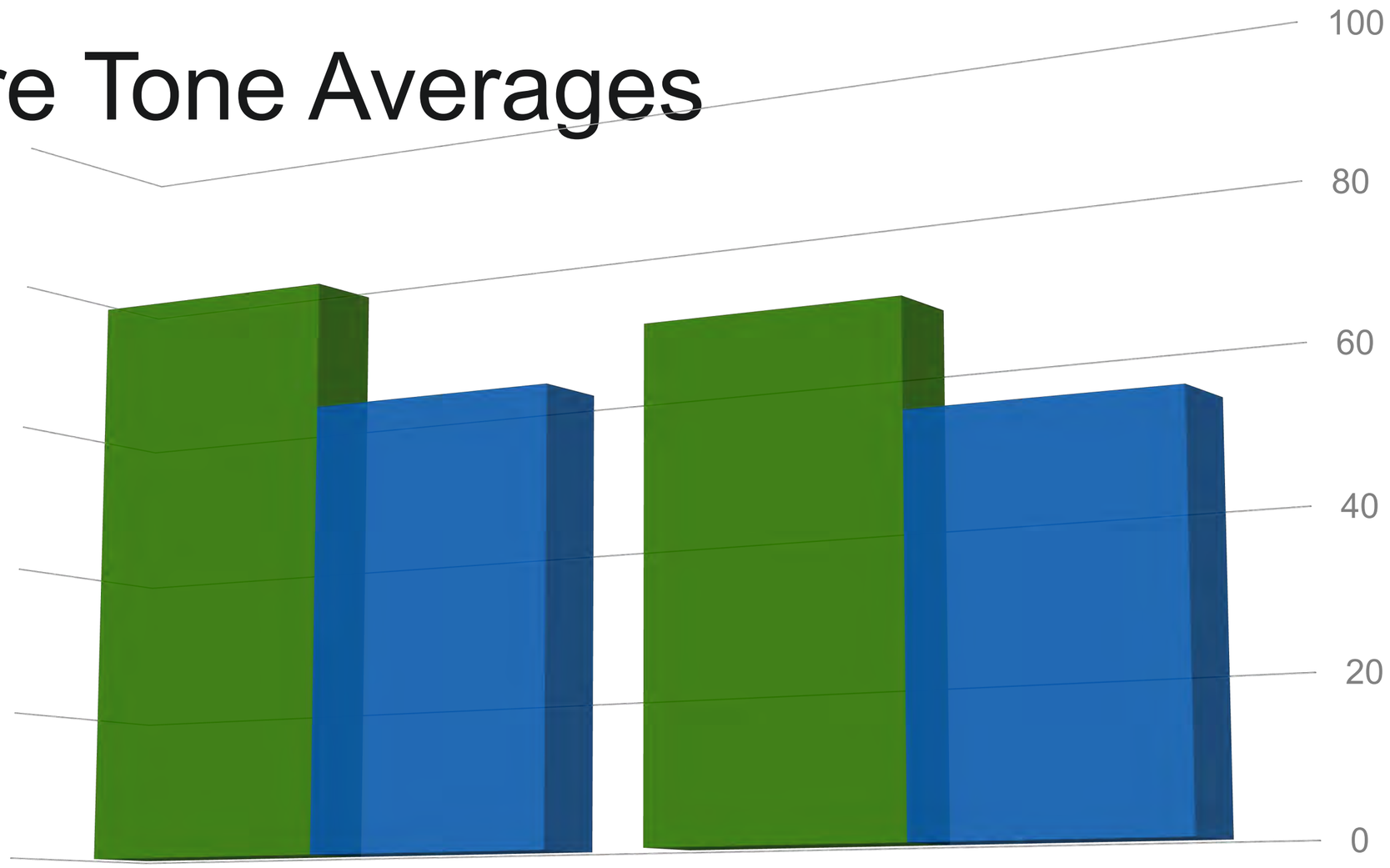
OPTION

Demographics

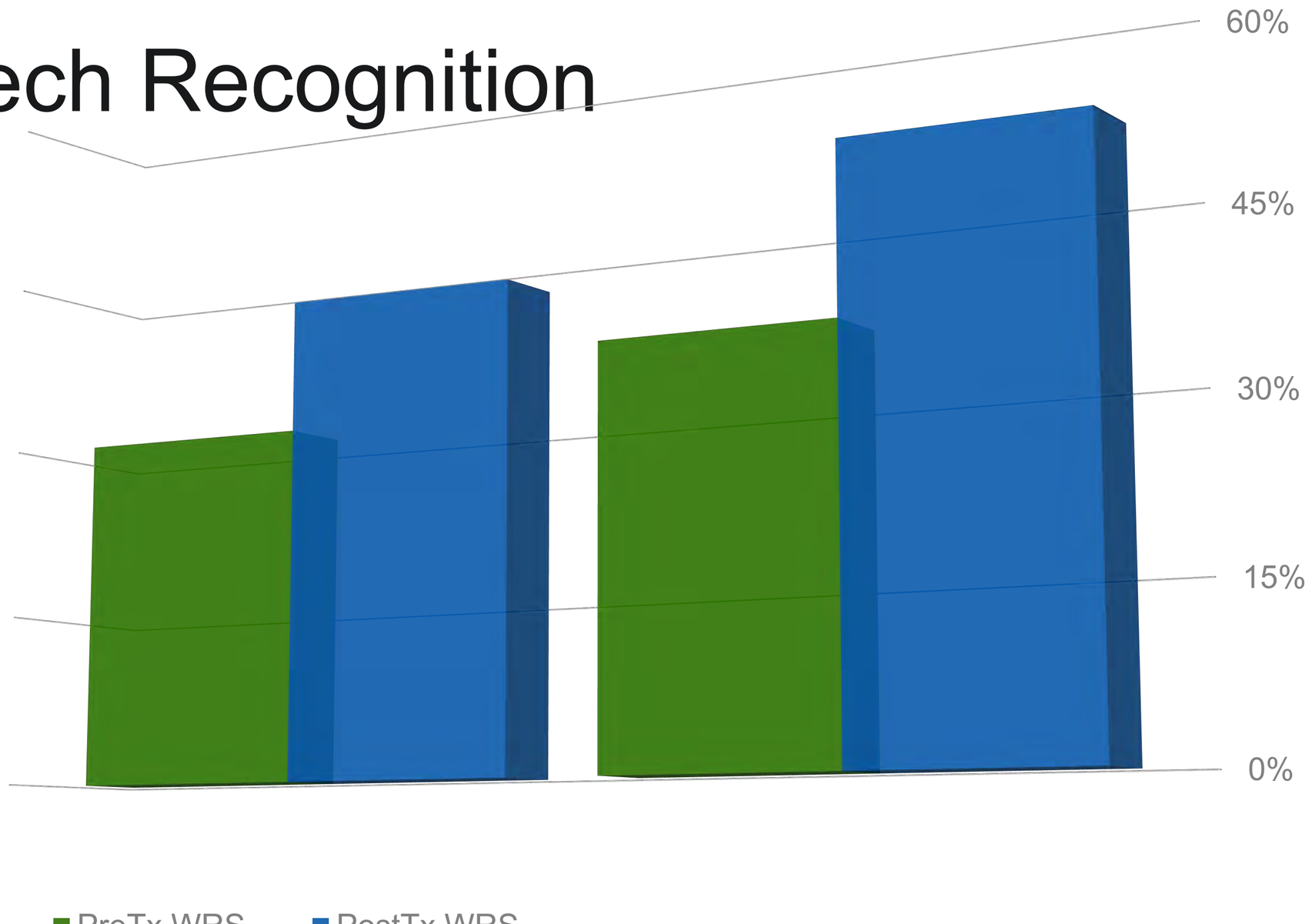
	Group 1 (N = 20)	Group 2 (N = 20)
Age (yrs)	54.7	58.2
Gender	9 Females	12 females
Pre-treatment PTA (dB)	77.4	66.9
Pre-treatment WRS (%)	30.8	36.2

*No significant differences between groups

Pure Tone Averages



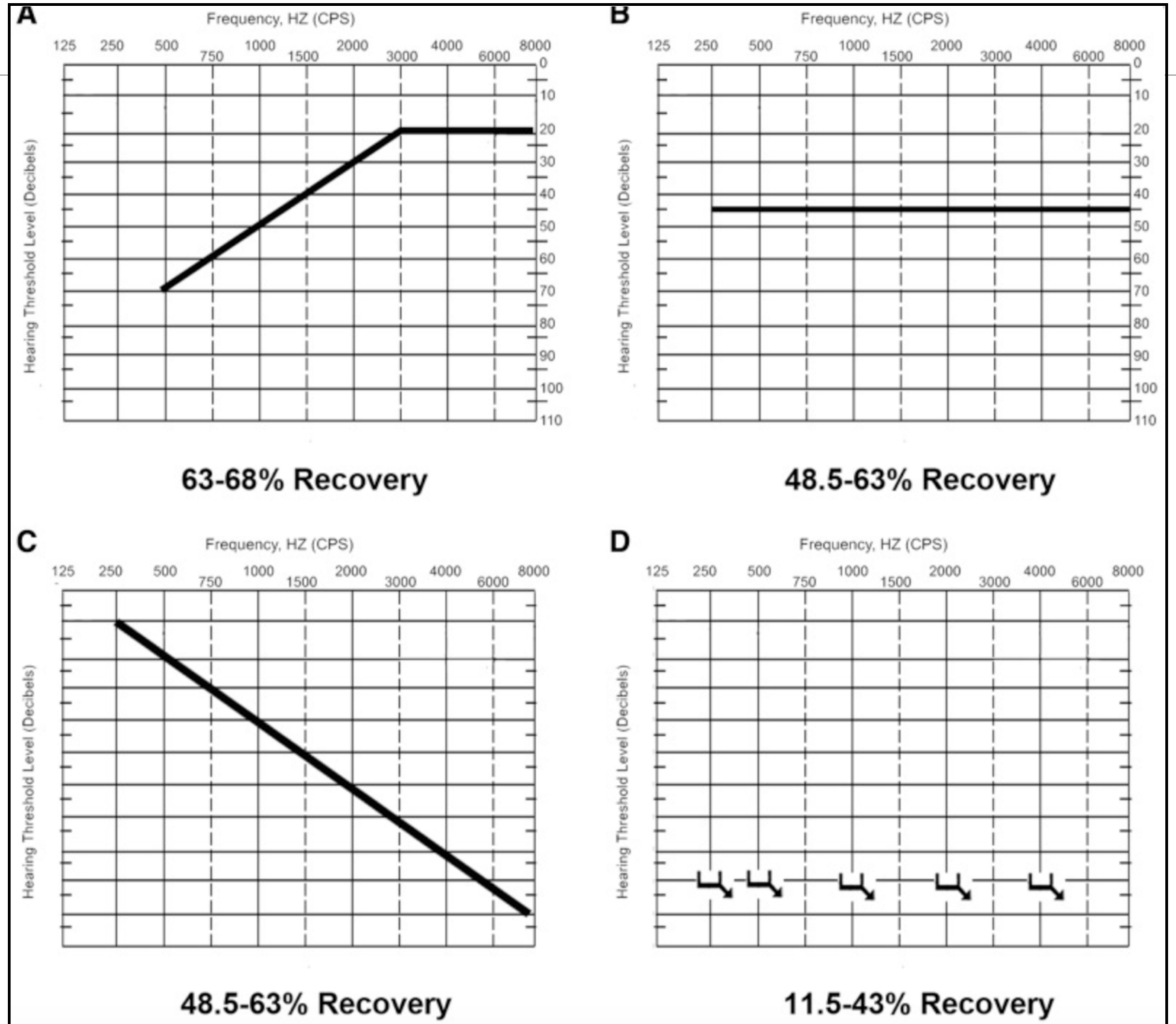
Speech Recognition

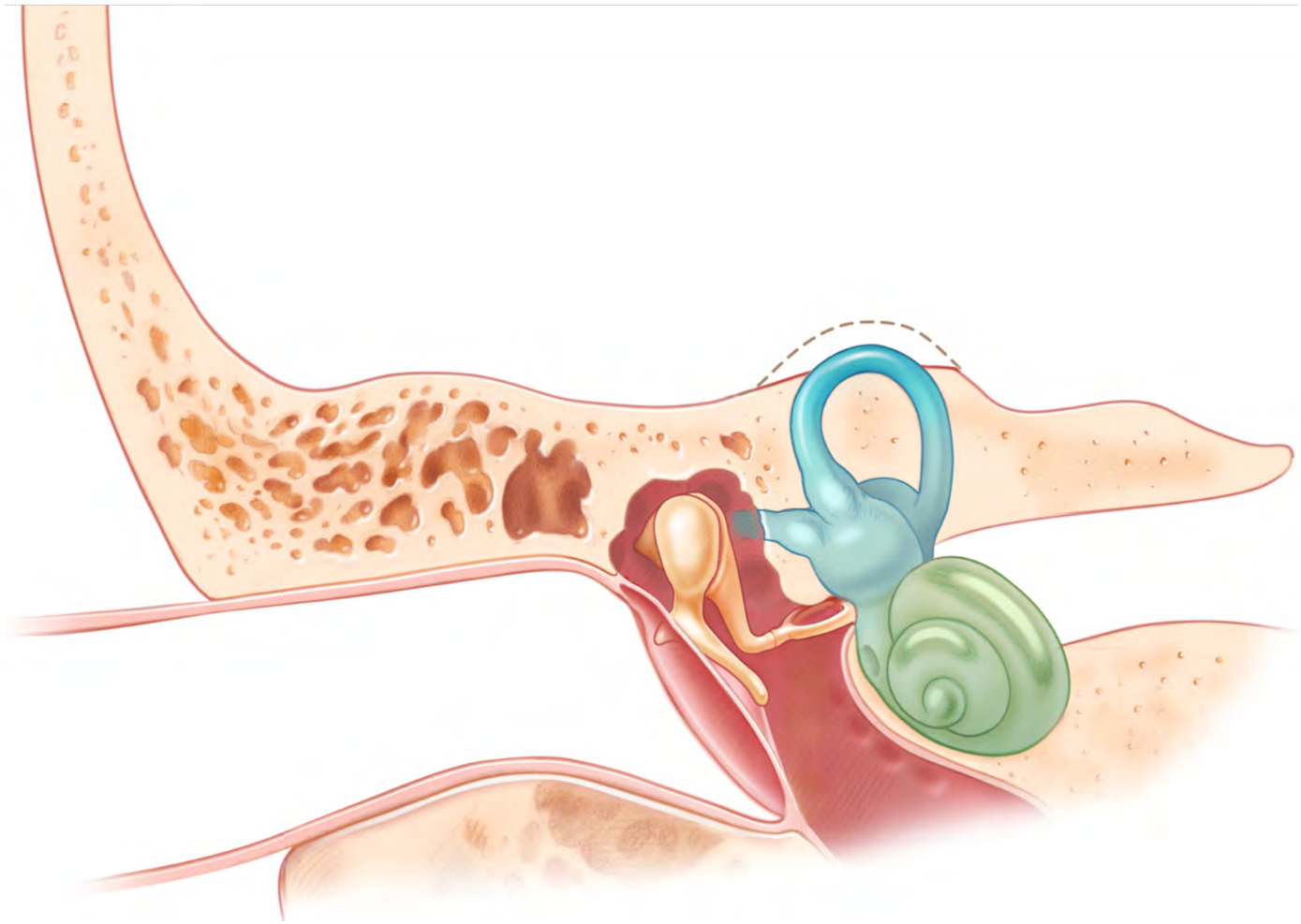


SSNHL: Prognosis

- Prognosis due to known etiology depends on the etiology and treatment options
- ISSNHL = 45-65% regain hearing (w/wo therapy), average gains of 35 dB
- Advanced age (> 60 years) correlates with poor prognosis
- Other factors: severity of loss, duration, vertigo, time to treatment, ‘shape’ of audiogram

SSNHL: Prognosis



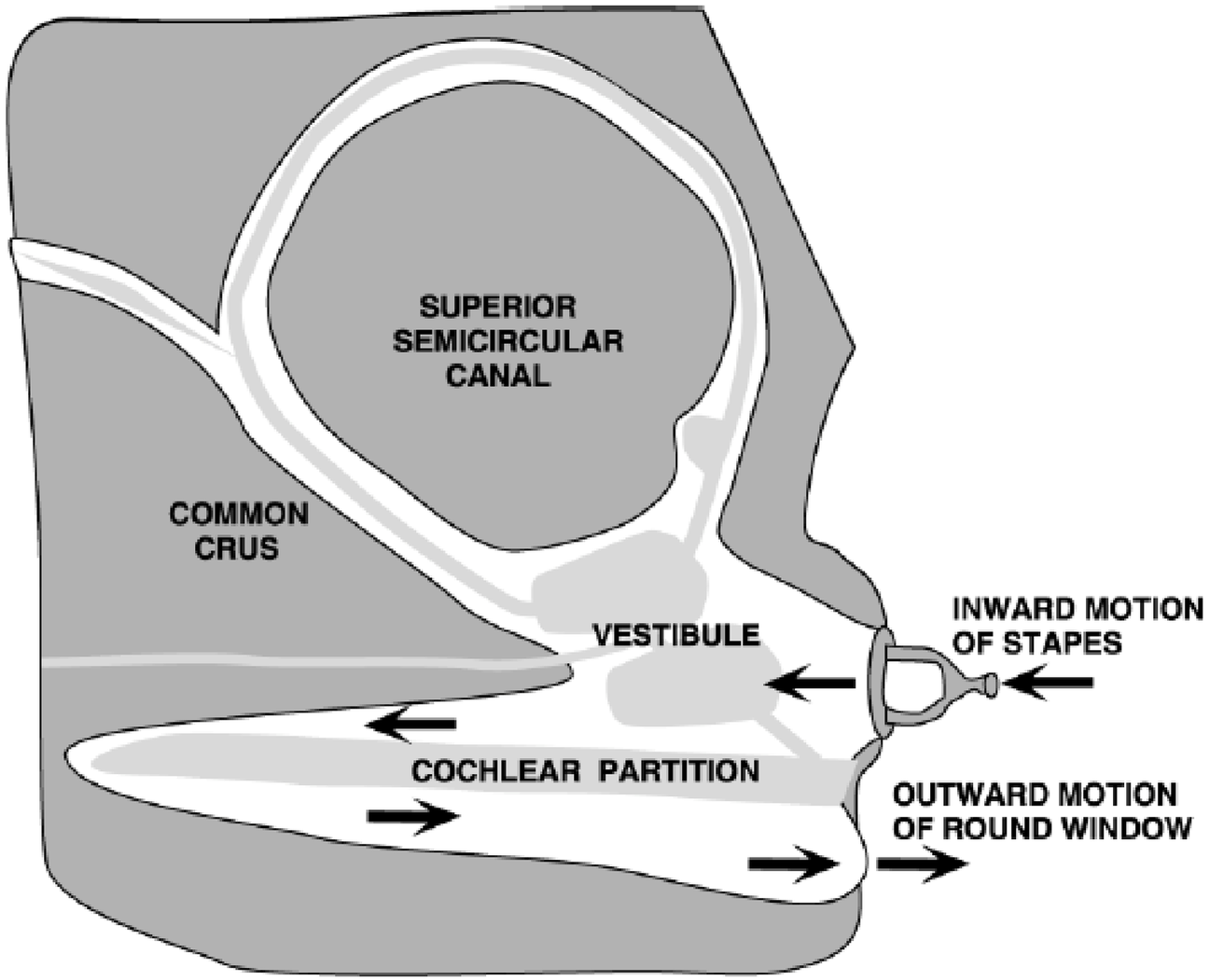


Superior Semicircular Canal Dehiscence Syndrome

SCDS: Background

- 1998 - series of patients with chronic disequilibrium, sound-induced vertigo, nystagmus in SSCC plane *(9525507)*
- Additional findings of BC hyperacusis and PT
- CT findings showed missing bone over the superior canal
- Etiology: congenital vs. acquired - congenital favored
- Temporal bone study: 0.5% SSCD (0.7% subjects) (1.4% near dehiscence) *(10680863)*
- Not all symptomatic

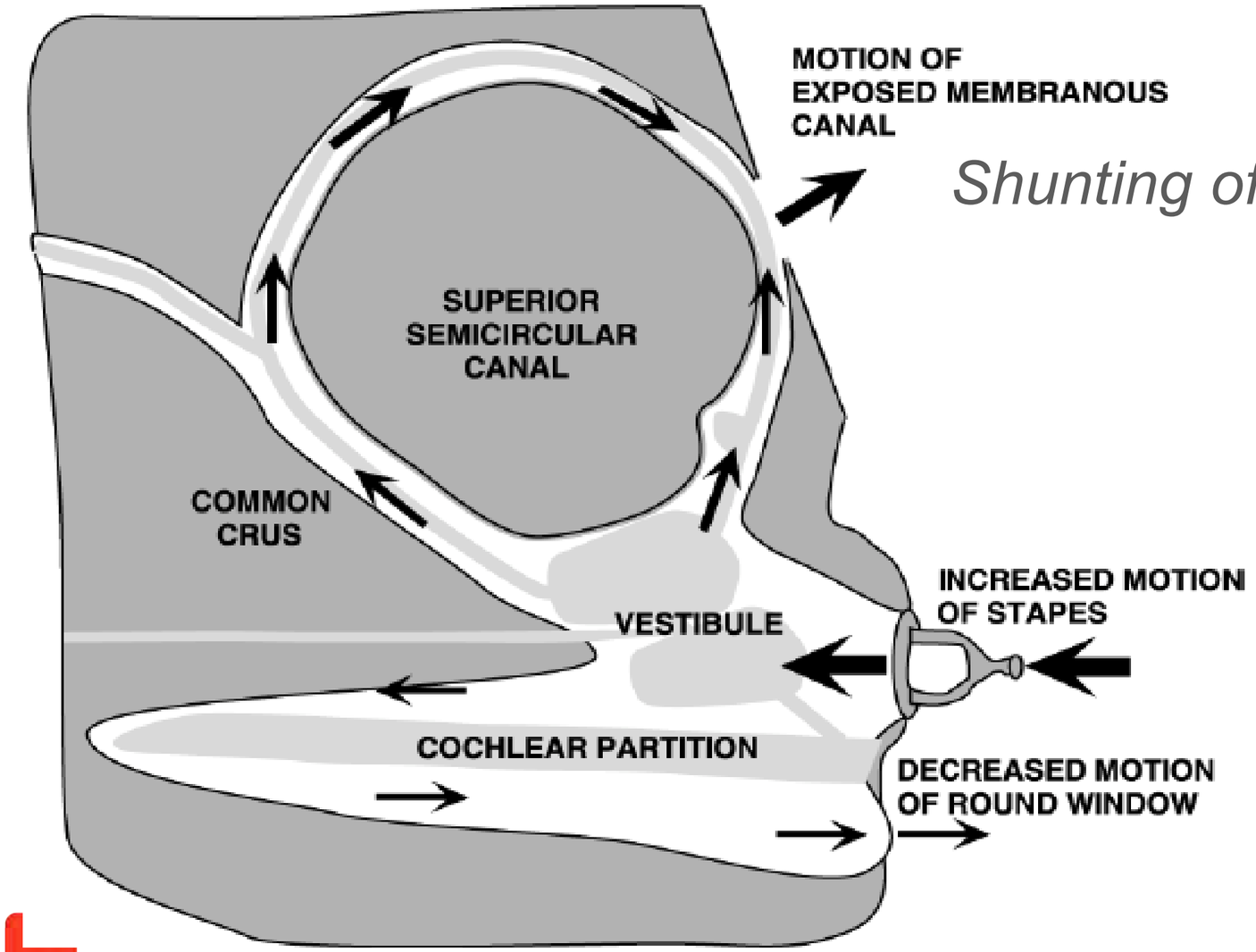
Hearing in a Normal Ear



Because inner ear is non-compressible, inward stapes motion = outward stapes motion

Chien et al, Otol Neurotol, 2007

Increased Air Conduction Threshold in SCDS

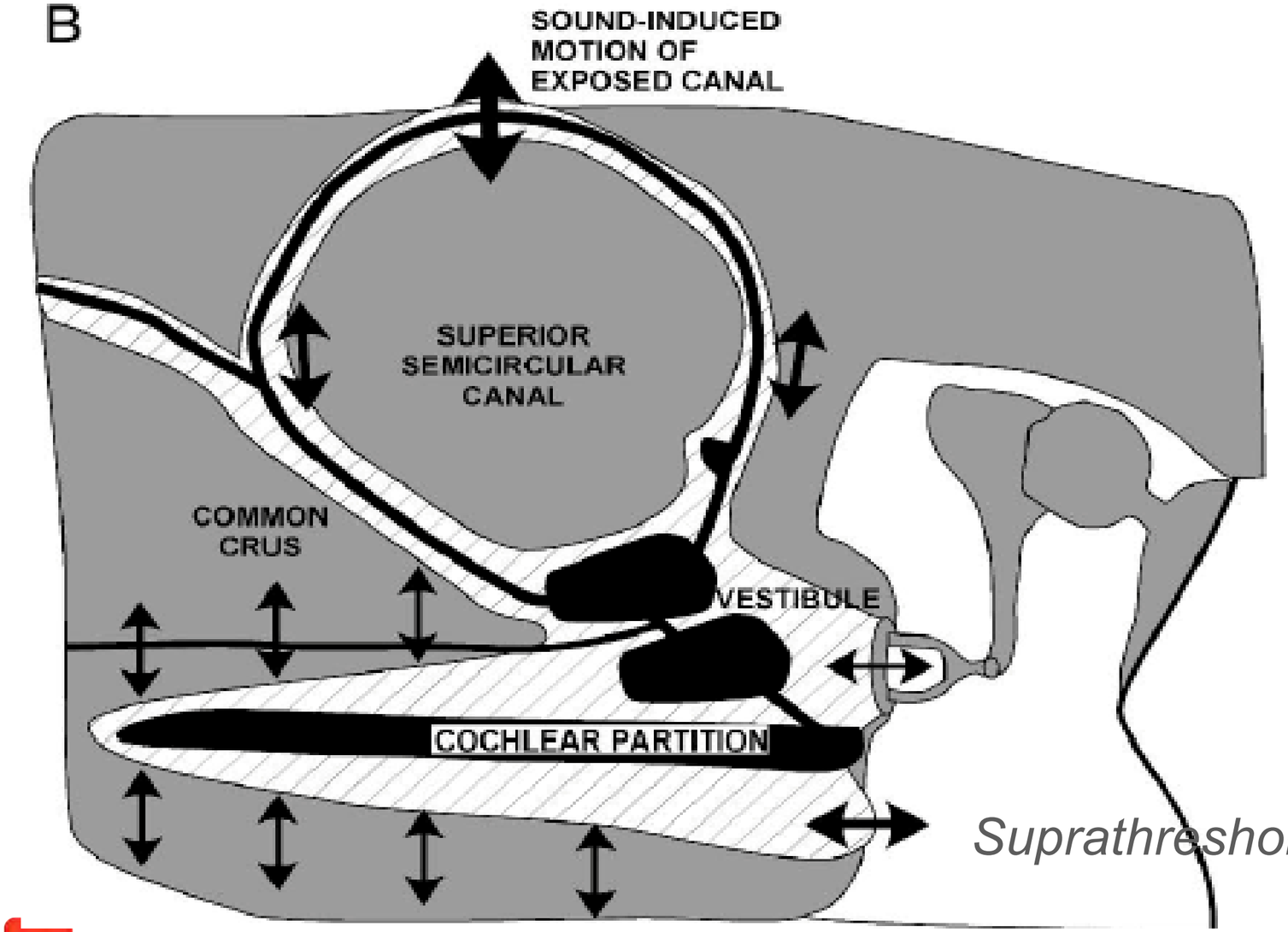


Shunting of fluid volume

Decreased round window motion

Chien et al, Otol Neurotol, 2007

Decreased Bone-Condition Thresholds



Increased response by cochlea to compressional wave

Suprathreshold fluid motions

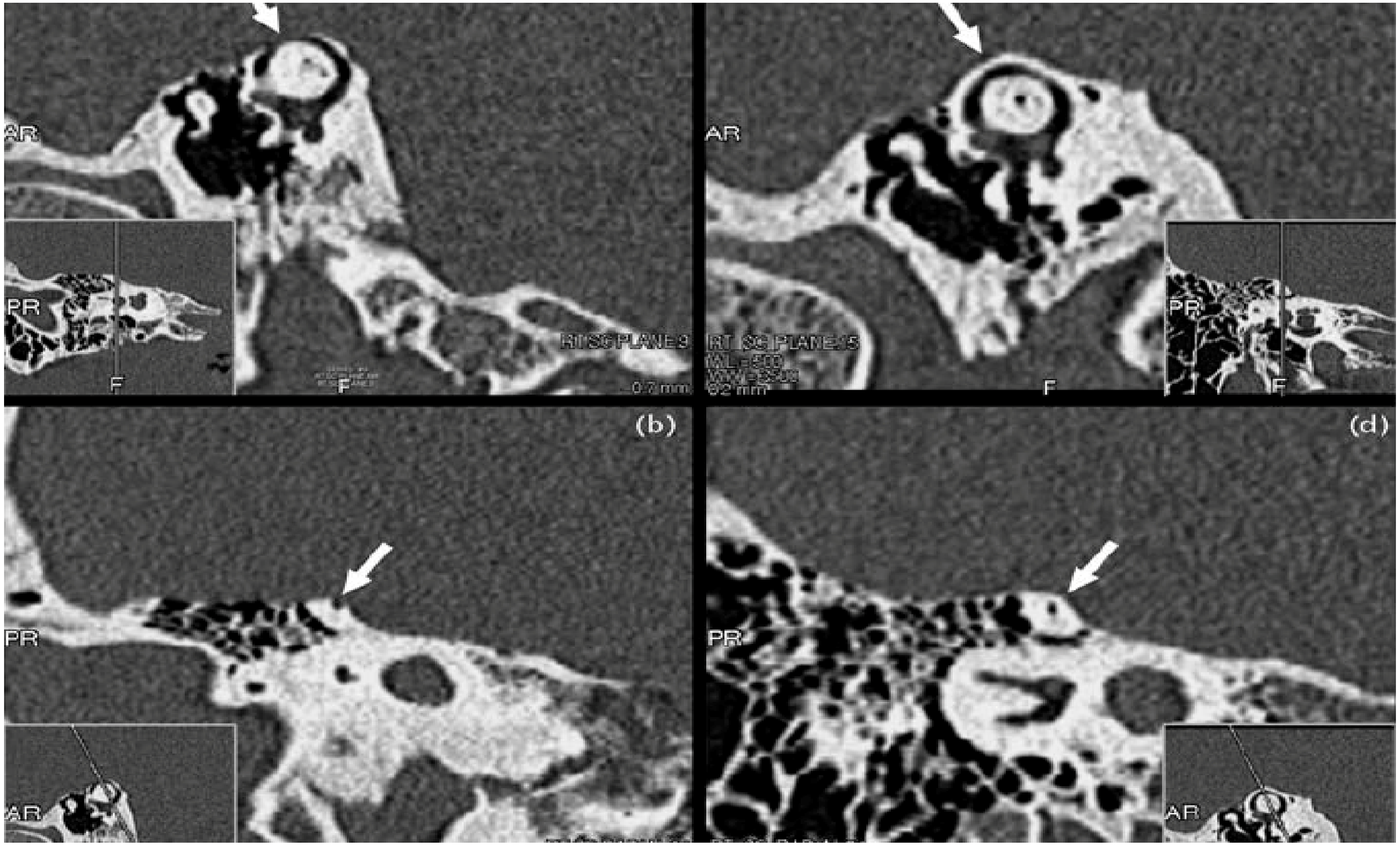
SCDS: Symptoms

- Superior canal dehiscence syndrome
  - BC hyperacusis, autophony, PT, sound/pressure induced vertigo
  - Conductive hearing loss - does not change over time
- Symptoms may worsen over time with progression of dehiscence size
- Size > 2.5 mm = vestibulocochlear symptoms vs. < 2.5 mm either vestibular or cochlear symptoms *(20118818)*
- No association between size and audiogram configuration although length may be correlated to ABG *(22664896)*

Pfammatter et al, Otol Neurotol, 2010

SCDS: Diagnosis

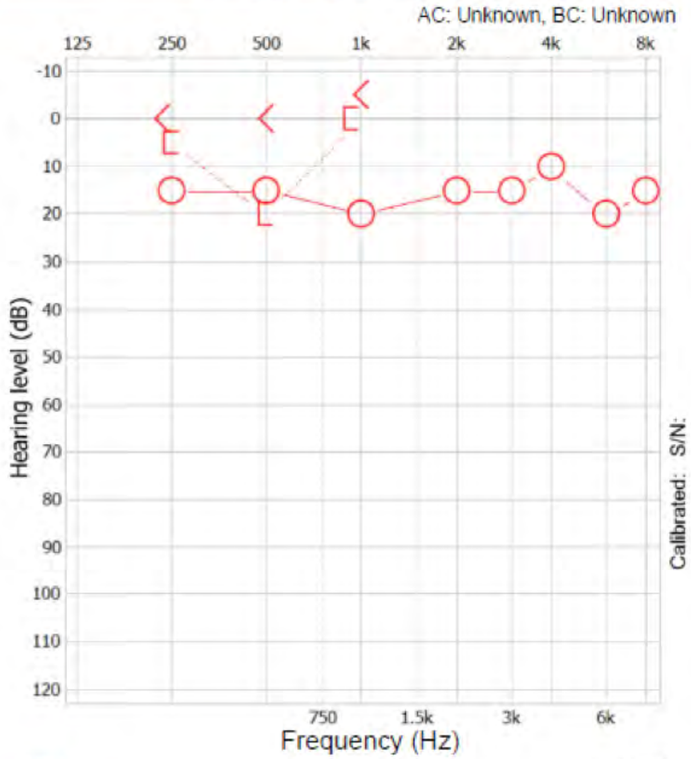
- Exam: Valsalva: downbeating nystagmus with torsional fast phase
- Audiology
  - ABG in lower frequencies, suprathreshold BC
  - ARTs commonly normal
- VEMP: reduced cVEMP thresholds (saccule), elevated oVEMP (utricle)
  - oVEMP highly sensitive and specific
- ECochG: elevated SP:AP ratio
- CT Temporal bone (thickness < 0.625, reformatted)


AR
PR
F
(b)
(d)

SCDS: Treatment

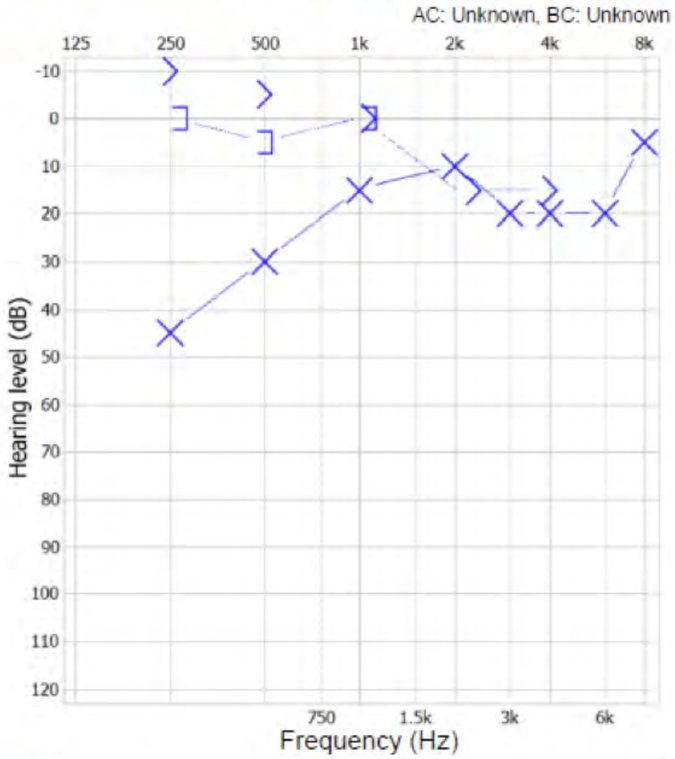
- Medical treatment symptomatic
- Less than 50% with known diagnosis pursue surgery
- Goal of surgery is to close 3rd window pathophysiology
- Middle fossa craniotomy vs. transmastoid
  - Patient anatomy, surgeon experience
  - VM experience - similar symptom control, higher rate of revision with MCF
  - Low risk of hearing loss, HF SNHL common (25%)

RIGHT (11/12/2014)



AC			
BC	100	70	70

LEFT (11/12/2014)



AC			
BC	75	60	65

PTA (dB HL) / AI (%)

	AC	BC	AI
Right	16		98
Left	18	6	86

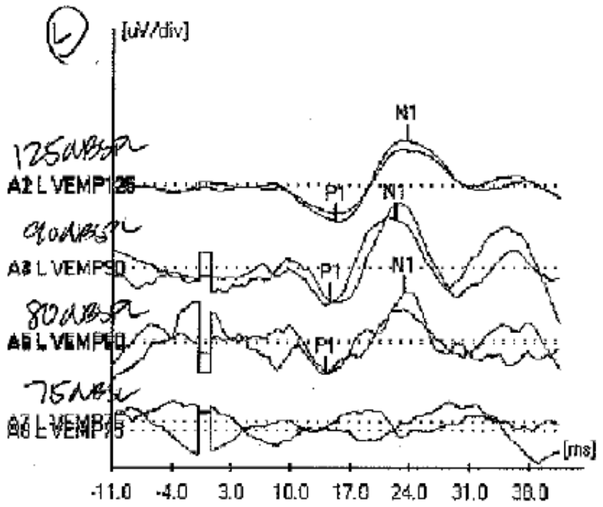
Reliability

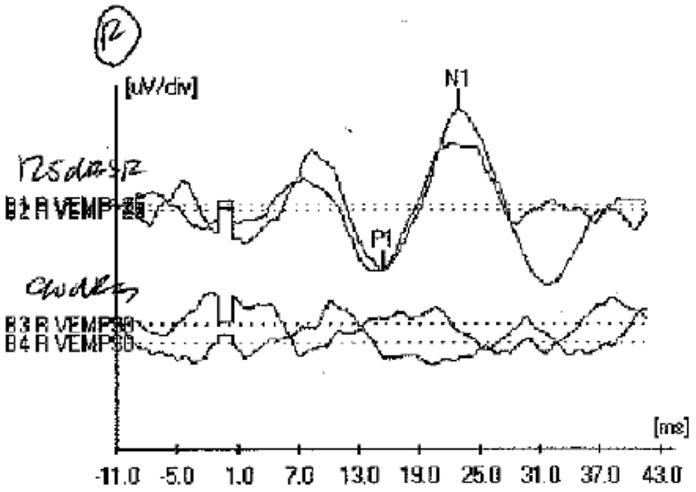
Speech	SDT		SRT		WRS / SRS 1				WRS / SRS 2				MCL	UCL
	dB HL	[m]	dB HL	[m]	%	dB HL	[m]	S/N	%	dB HL	[m]	S/N	dB HL	dB HL
Right			20		96	60	[55]							
Left			20		100	60	[55]							

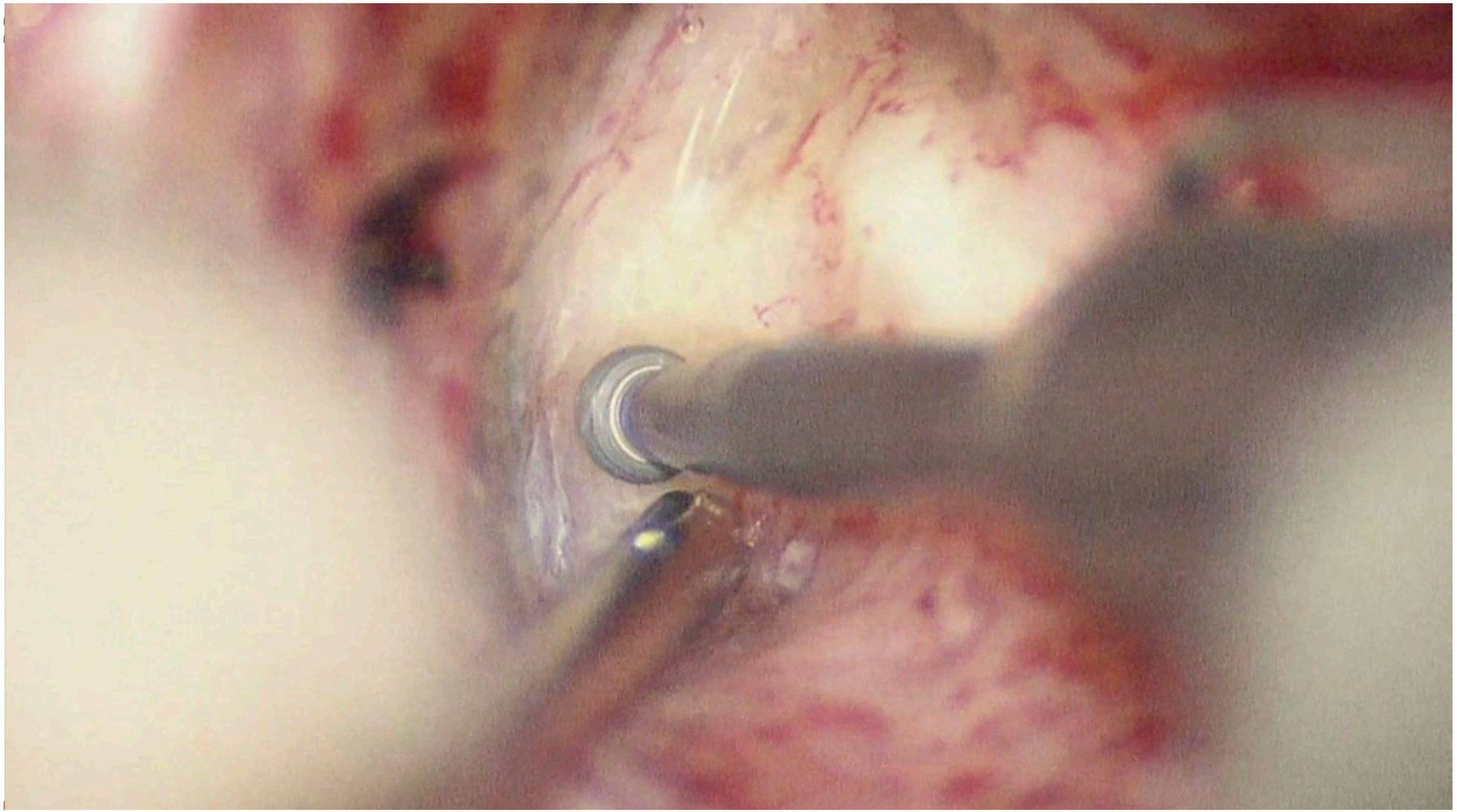
Legend

L	R	Masked
×	○	AC □ △
>	<	BC ⌋ ⌊
S	S	SF ⊗ ∅
M	M	MCL
U	U	UCL
»	*	NR

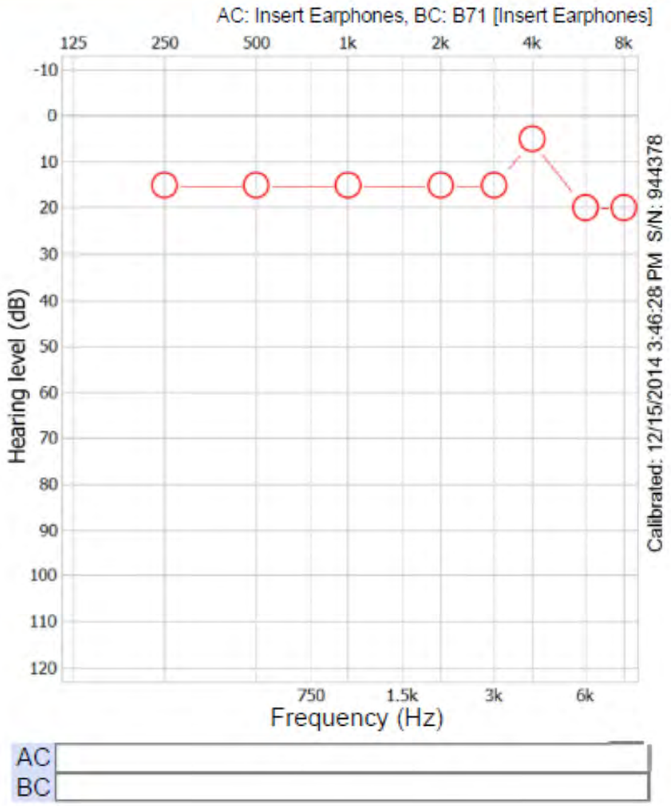
PTA: AC: 500, 1k, 2k
BC: 500, 1k, 2k





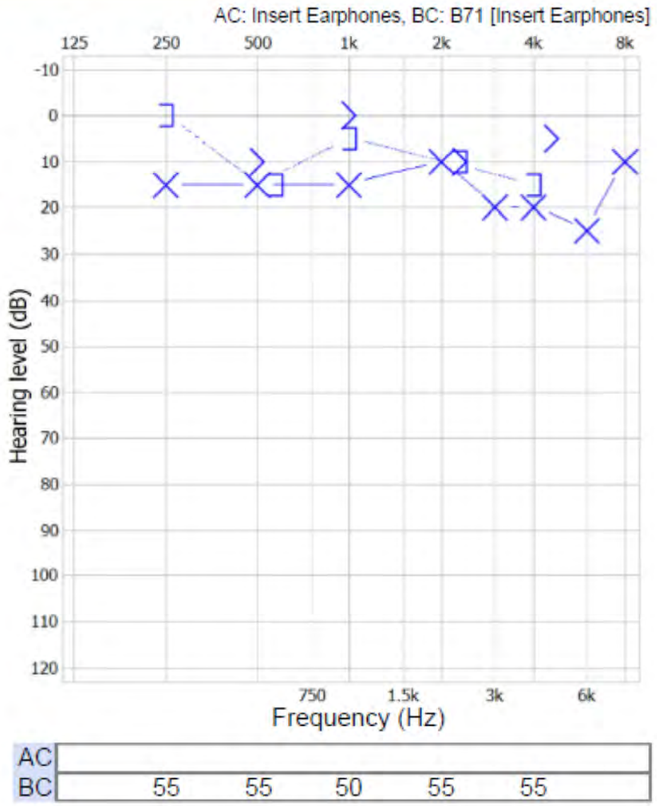


RIGHT (2/11/2015)



AC	
BC	

LEFT (2/11/2015)



AC					
BC	55	55	50	55	55

PTA (dB HL) / AI (%)

	AC	BC	AI
Right	15		98
Left	13	10	95

Reliability

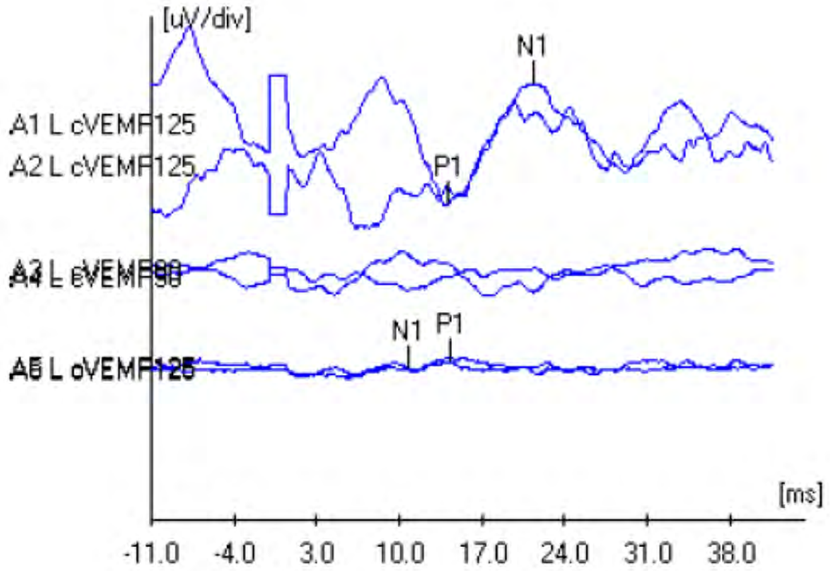
Good

Legend

L	R	Masked
	AC	
	BC	
S	SF	
M	MCL	
U	UCL	
	NR	

PTA: AC: 500, 1k, 2k
BC: 500, 1k, 2k

Speech	SDT		SRT		WRS / SRS 1				WRS / SRS 2				MCL	UCL
	dB HL	[m]	dB HL	[m]	%	dB HL	[m]	S/N	%	dB HL	[m]	S/N	dB HL	dB HL
Right			15		100	55								
Left			15		100	55								
Bin														



Hearing Outcomes Following SSCD Repair

- SCD repair NOT a hearing loss surgery
  - Ward: 43 ears, ABG 16 dB — 8 dB; 53% increased PTA (8 dB-19 dB) *(22935810)*
  - Limb: 19 primary ears, no change AC or BC, partial closure of ABG in 5/19 *(17006348)*
  - Goddard: 24 ears, no difference in AC thresholds *(24962178)*
  - Yuen: 10 ears, 1 complete ABG closure, 6 partial, 3 increased *(19326497)*

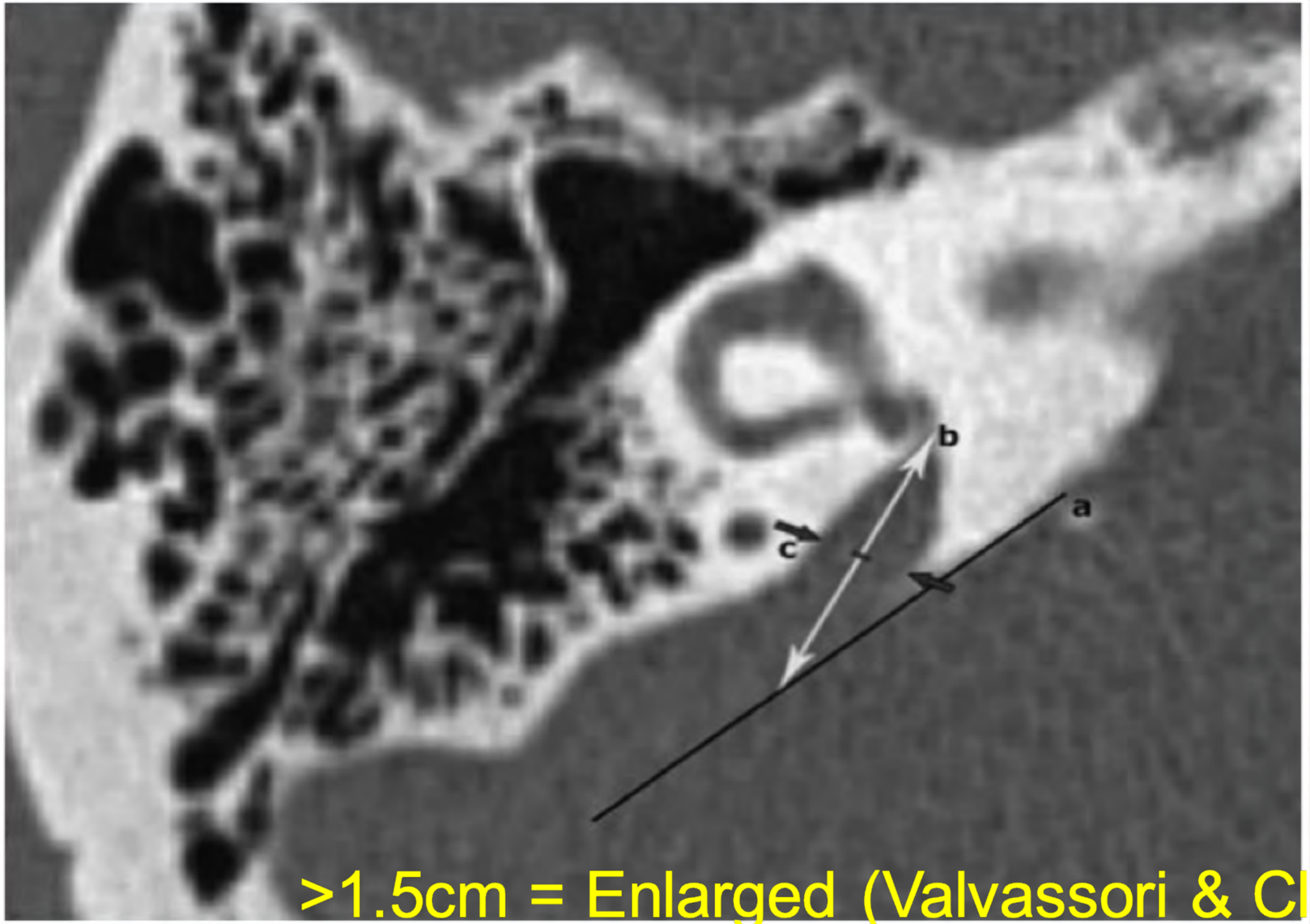
Enlarged Vestibular Aqueduct (Large Vestibular Aqueduct Syndrome)

LVAS

- Enlargement of bony aqueduct that contains endolymphatic duct
- Most common radiographic abnormality in people with SNHL (? cause of HL)
- HL resulting from pressure fluctuations in inner ear
- Genetic disorders: Pendred, Waardenburg's, X-linked stapes gusher syndrome, branchio-oto-renal syndrome

LVAS: Diagnosis

- Audiometry: fluctuating SNHL, often with MHL
  - Mixed component thought due to 3rd window
- Vestibular testing not helpful
- CT and MRI allow definitive diagnosis



>1.5cm = Enlarged (Valvassori & Clemis)

Head Trauma and EVAS

- Noordman BJ et al, *Otol Neurotol,* 2014 (25406869)
  - 31 studies, 179 patients, 351 EVAs
  - 34% SHL after head/noise/barotrauma
  - Pre-trauma fluctuating hearing correlates with SHL after trauma (OR 8.6)
  - VA diameter, type of preexisting HL, severity of HL, preexisting progressive HL, and diagnosis of Pendred not associated

Thank you

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