Ménière's Disease and Surgery for Intractable Vertigo

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Ménière's Disease

- Prosper Ménière:
  - 1861
  - first report of a group of patients with disabling attacks of vertigo with associated hearing loss and aural symptoms

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Ménière's Disease

- PREVALENCE
  - 3.5-500/100,000 (worldwide)
  - 200/100,000 (U.S.)
- INCIDENCE
  - 8-150/100,000/year (worldwide)
  - 15/100,000/year (U.S.) ~ 50,000/yr

By Comparison:	
Allergies:	17,000 /100,000
Hearing Loss:	8,000-17,000 /100,000
Rheumatoid Arthritis:	600/100,000
Multiple Sclerosis:	22-160/100,000
Acoustic Neuroma:	1/100,000/year

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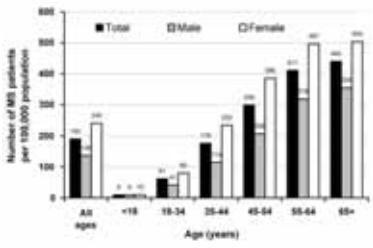
Ménière's Disease - Epidemiology

- GENDER
  - F:M 1.5:1 to 4:1 (worldwide)
  - F:M 1.9:1 (U.S.)
- AGE
  - 8-150/100,000/year (worldwide)
  - <18yrs: 9/100,000 (U.S.)
  - >65yrs: 440/100,000 (U.S.)



Alexander TH, *Otolaryngol Clin North Am*, 2015

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Unilateral v Bilateral

- BILATERAL DISEASE: 15-50% reported
  - most studies: 15-25%
- 90% of latent contralateral ears present with S/Sx (HL, aural Sx) within 5 years of presentation of initial ipsilateral ear
- >90% of the time, the HL in latent contralateral ears remains less severe than HL in initial ipsilateral ear

Palaskas CW, Dobie RA, Snyder JM, *Laryngoscope*, 1988

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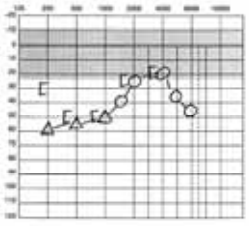
Ménière's Disease - Diagnosis

CLINICAL TETRAD

- HEARING LOSS
  - usually low frequency
- TINNITUS
  - often lower pitched
  - buzzing, machinery, ocean
- AURAL FULLNESS
- EPISODIC VERTIGO
  - typically minutes - hours
  - not seconds, not "days" - but be careful of description



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Ménière's Disease - Testing

- Audiometry
- Glycerol Dehydration Test
- ECoG
- ENG
- VEMP
- 68 kD / HSP-70 Western Blot

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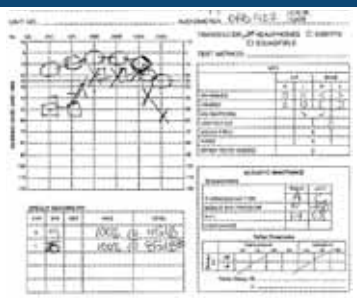
Audiometry



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Glycerol Dehydration Test

- Pre-ingestion audiogram ->
  - Glycerol (or mannitol) 1.5g/kg (1.2cc/kg) PO ->
    - Wait 1-2 hrs ->
      - Post-ingestion audiogram
- (+) Test for MD:
  - ↑ ≥ 10dB in 2+ frequencies or ↑ ≥ 12% in SDS
- Specificity 90%, Sensitivity 40-60%
- Test is old and outdated - but mostly unpleasant and impractical - rarely used

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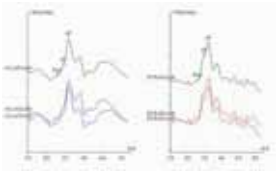
ECoG

Electrocochleography



- SP/AP ratio:
  - abnormal: >0.45-0.50
  - MD sensitivity 70%
  - MD specificity 90+%
- requires placement of promontory electrodes: uncomfortable, impractical

Chung et al. 2004. *O&N.* 25:144-9.

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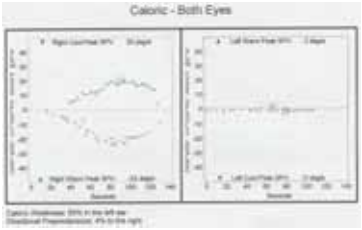
ENG

Electronystagmography

- UW: Abnormal > 25% (20%, 30%)
- DP: Abnormal > 30% (20%)
- sensitivity for MD poor
  - typically normal unless having active attack



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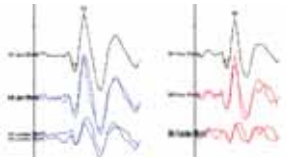
cVEMP

Cervical Vestibular Evoked Myogenic Potential P1/N1

- P1/N1
  - Threshold: NL 85-90dB
  - (click)
    - ↑ in MD (≥110dB)
    - ↓ in SCDS (≤70dB)
  - Amplitude: NL ≥ 70uV
    - ↓ in MD
  - sensitivity for MD poor ~60%
  - not used for Dx



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HSP-70 / 68kDa Western Blot

- Test developed to assess responsiveness to steroids in cases of AIIED
- Poor sensitivity/specificity
- Some with MD are (+) (~10-20%)
- => Worthless test (in this indication)

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TESTING SPECIFICITY

- Audiometry
- Glycerol Dehydration Test
- ECoG
- ENG
- VEMP
- 68 kD / HSP-70 Western Blot

MENIERE'S DISEASE IS A CLINICAL DIAGNOSIS

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Definition

AAO-HNS Committee on Hearing and Equilibrium (rev. 1995)

Table 1. Diagnosis of Meniere's disease

Certain Meniere's disease	DEAD
Definite Meniere's disease, plus histopathologic confirmation	
Definite Meniere's disease	
Two or more definitive spontaneous episodes of vertigo, 20 minutes or longer	2 + HL
Audiometrically documented hearing loss on at least one occasion	
Tinnitus or aural fullness in the treated ear	
Other causes excluded	
Probable Meniere's disease	1 + HL
One definitive episode of vertigo	
Audiometrically documented hearing loss on at least one occasion	
Tinnitus or aural fullness in the treated ear	
Other causes excluded	
Possible Meniere's disease	Vertigo or HL
Episodic vertigo of the Meniere type without documented hearing loss, or	
Sensorineural hearing loss, fluctuating or fixed, with dysequilibrium but without definitive episodes	
Other causes excluded	

Otolaryngology - Head and Neck Surgery, 1995; 113:181-5.

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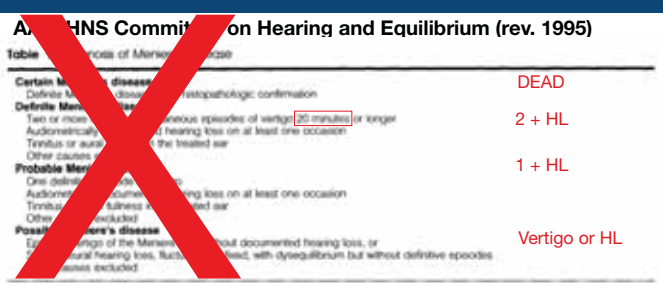
Definition

AAO-HNS Committee on Hearing and Equilibrium (rev. 1995)



Otolaryngology - Head and Neck Surgery, 1995; 113:181-5.

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Newer Diagnostic Criteria

International Consortium of Otolaryngology Societies

- Barany Society
- Japan Society for Equilibrium Research
- European Academy of Otology and Neurotology
- AAO-HNS CHE
- specifies hearing loss as "low- to medium-frequency"
- specifies vertigo episode duration as 20min-12hrs (definite) or 20min-24hr (probable)
- illustrates difficulty in differentiating MD from other emerging entities of recurrent vertigo (MRV, BRV+SNHL)

Lopez-Escamez JA et al., *Acta Otorrinolaringol Esp*, 2015

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Definition

AAO-HNS Equilibrium Committee (rev. 2016)

Table 2. Amended 2015 Criteria for Diagnosis of Ménière's Disease

Definite	Two or more spontaneous episodes of vertigo, each lasting 20 min to 12 h	
	Audiometrically documented low- to midfrequency sensorineural hearing loss in 1 ear, defining the affected ear on at least 1 occasion before, during, or after 1 of the episodes of vertigo	2 + HL
	Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear	
	Not better accounted for by another vestibular diagnosis	
Probable	Two or more episodes of vertigo or dizziness, each lasting 20 min to 24 h	Vertigo but no documented HL
	Fluctuating aural symptoms (hearing, tinnitus, or fullness) in the affected ear	
	Not better accounted for by another vestibular diagnosis	
Possible Ménière's disease		
Episodic vertigo of the Ménière type without documented hearing loss, or		
Sensorineural hearing loss, fluctuating or fixed, with dysequilibrium but without definitive episodes		
Other causes excluded		

Otolaryngology - Head and Neck Surgery, 2020; 162(2S):S1-S55.

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"Official" Staging of MD

for Certain, Definite, and Probable MD

AAO-HNS C.H.E. GUIDELINES FOR MENIERE'S DISEASE STAGING *	
Stage	**4 Tone Average (dB HL)**
1	≤ 25
2	26 – 40
3	41 – 70
4	> 70

Otolaryngology - Head and Neck Surgery, 1995; 113:181-5.

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"Official" Scaling of Functional Level

Patient must choose single level that best suits symptoms

Table 3. Functional level scale

Regarding my current state of overall function, not just during attacks (check the ONE that best applies):

1. My dizziness has no effect on my activities at all.
2. When I am dizzy I have to stop what I am doing for a while, but it soon passes and I can resume activities. I continue to work, drive, and engage in any activity I choose without restriction. I have not changed any plans or activities to accommodate my dizziness.
3. When I am dizzy I have to stop what I am doing for a while, but it does pass and I can resume activities. I continue to work, drive, and engage in most activities I choose, but I have had to change some plans and make some allowance for my dizziness.
4. I am able to work, drive, travel, take care of a family, or engage in most essential activities, but it must exert a great deal of effort to do so. I must constantly make adjustments in my activities and budget my energies. I am barely making it.
5. I am unable to work, drive, or take care of a family. I am unable to do most of the active things that I used to. Even essential activities must be limited. I am disabled.
6. I have been disabled for 1 year or longer and/or I receive compensation (money) because of my dizziness or balance problem.

Otolaryngology - Head and Neck Surgery, 1995; 113:181-5.

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Measuring and Reporting Treatment Outcomes

- OBJECTIVE
  - frequency of attacks
    - CHE 1995 guidelines - vertigo frequency
  - audiometry
    - CHE 1995 guidelines - audiometric staging

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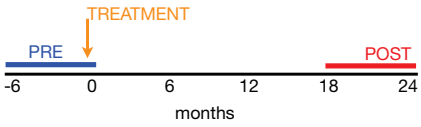
Treatment Outcomes

AAO-HNS Committee on Hearing and Equilibrium (rev. 1995)

Categorizes success of treatment based on differential improvement in vertigo frequency 6 months pre-treatment v. 18-24 months post-treatment



Otolaryngology - Head and Neck Surgery, 1995; 113:181-5.

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Measuring Treatment Outcomes

- SUBJECTIVE
  - patient questionnaires - many variations
  - DHI, THI, HHI
  - MD-POSI - Patient Oriented Severity Index UW
  - MDOQ - MD Outcomes Questionnaire MEI
- QOL studies gaining in importance/popularity
  - 1 prior to 2004, 8 from 2004-2015
- 143 clinical studies on MD 2004-2015

Syed MI et al., *Otol Neurotol*, 2015

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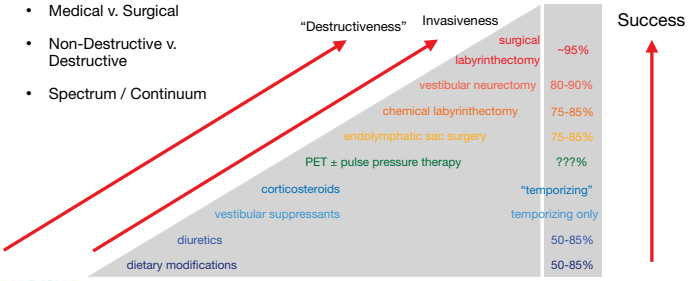
Treatment Options

- Medical v. Surgical
- Non-Destructive v. Destructive
- Spectrum / Continuum



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FIRST ECHELON TREATMENTS

- DIETARY MODIFICATIONS
- DIURETICS
- VESTIBULAR SUPPRESSANTS
- SYSTEMIC STEROIDS
- INTRATYMPANIC STEROIDS

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Dietary Modifications

- average American diet 5000 - 7000 mg / day
- Recommendations for MD patients:
  - < 1500 mg / day
  - (same as AHA recommendations for Htn, CAD)

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Diuretics

- HCTZ
- Triamterene
- combinations: Dyazide, Maxzide
- Aldactone
- acetazolamide / Diamox

- Escalating therapy
- Sulfa allergy
- Electrolyte monitoring and K supplementation

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Diet + Diuretic

- Daily diuretic (any) + dietary modifications (salt restriction<1500mg/day) leads to improvement in MAJORITY of MD patients
- Continuous diuretic therapy with dietary modifications 24+mo:
- Vertigo:
  - 79% - CHE class A/B/C (substantial to complete improvement)
  - 19% - CHE class D (limited to no improvement)
  - 2% - CHE class E (worse)
- Hearing:
  - 1/3 improved / 1/3 unchanged / 1/3 worse

Santos PM et al., *Otolaryngol Head Neck Surg*, 1993

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Photosensitizing Diuretics and Skin Cancer

Photoactivation leads to ROS, damage to cell membranes & DNA

- Photosensitivity/Phototoxicity of diuretics (and oral antidiabetics) well documented for decades - Stern RS et al., *NEJM*, 1984; Selvaag H et al., *In Vivo*, 1997; Stern RS et al., *J Natl Cancer Inst*, 1998.
- First large population based survey demonstrating association of photosensitizing diuretics with skin cancer - Jensen AØ et al., *British J Cancer*, 2008
  - SCCA: amiloride+HCTZ - IRR 1.79 (95%CI 1.45-2.20)
  - MM: indapamide - IRR 3.30 (95%CI 1.34-8.10)

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Photosensitizing Diuretics and Skin Cancer

Balancing Risk & Benefit

- LANDMARK DANISH CANCER REGISTRY POPULATION BASED STUDIES:
  - Pedersen SA et al., *J Am Acad Dermatol*, 2018 - [NMSC]
  - Pottegard A et al., *JAMA Intern Med*, 2018 - [MM]

DOSE	HISTOLOGY	ODDS RATIO	95% CI
HCTZ ≥ 50,000mg	BCCA	1.29	(1.23-1.35)
	SCCA	3.98	(3.68-4.31)
	MM - total	1.22	(1.09-1.36) no dose response
	MM - nodular	2.05	(1.54-2.72) P for trend = 0.01
	MM - lentigo	1.61	(1.03-2.50) P for trend = 0.16
	MM - superficial spreading	1.11	(0.97-1.27) P for trend = 0.73
HCTZ ≥ 100,000mg	MM - total	1.26	(1.08-1.46) P for trend = 0.24
HCTZ ≥ 200,000mg	BCCA	1.54	(1.38-1.71)
	SCCA	7.38	(6.32-8.60)

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Vestibular Suppressants

- antihistamines
- meclizine / Bonine / Antivert
- lorazepam / Ativan
- diazepam / Valium

- dosing: valium 2mg v. 5mg

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Corticosteroids

- Some causes of MD may be immune related
- Systemic vs Intratympanic delivery

Tomoda K et al., *Acta Otolaryngol*, 1993
Shea JJ, *Adv Otorhinolaryngol*, 1983
Hughes GB et al., *Laryngoscope*, 1983

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Systemic Steroids

- PO steroids: gold standard
  - ~25% responders
- Medrol Dose Pack - 6 days
- Prednisone - 10 days, 14 days (with taper)
- Systemic steroid risks

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Intratympanic Steroids

- Appear to be equally effective to PO steroids in some studies
- less practical: require patients to come in
  - daily x 5-7 days
  - weekly x 3-6 weeks
  - any other (impractical) combination you can think of
- Systemic risks traded for: TM perforation (10%)
- Useful for those where PO steroids are not tolerated or contraindicated, otherwise no added benefit

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SECOND ECHELON TREATMENTS

- PET TUBE ± PULSE PRESSURE THERAPY
- INTRATYMPANIC GENTAMICIN
- ENDOLYMPHATIC SAC SURGERY

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PET ± Pulse Pressure Therapy

- Pulse Pressure Therapy + PET - “Meniett Device”
  - mechanism of intervention not understood
- 5 clinical trials of Meniett + PET
  - 1/5 show benefit
- Pressure Equalization Tube (PET) alone may be beneficial
  - Sugawara K et al., *Auris Nasus Larynx*, 2003.
  - Ogawa Y et al., *J Laryngol Otol*, 2015.

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Intratympanic Gentamicin

"Chemical Labyrinthectomy"

- Many Treatment Protocols described
  - Titration
  - Fixed - seem to have slightly higher response rates
- 70-90% success rate in vertigo control (CHE class A/B)
- Seem to have slightly increased risk of HL - debated
- 2 prospective, double blinded, randomized, placebo controlled trials in past decade, none before 2004
  - results similar as prior studies

Wasson J, Upile N, Pfleiderer A, *J Laryngol Otol*, 2013
Stokroos R, Kingma H, *Acta Otolaryngol*, 2004
Cohen-Kerem R et al., *Laryngoscope*, 2004

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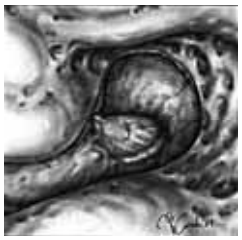
Endolymphatic Sac Surgery

"Enhancement" Surgery

- Endolymphatic Sac Shunt or Drainage
- Endolymphatic Sac (± Vein) Decompression



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Endolymphatic Sac Surgery

- EFFECTIVENESS: (CHE class A/B/C)
  - 75% - short term < 1 yr
  - 75-85% - long term > 2 yrs
  - hearing either stable or potentially can improve in short term
  - QOL outcomes, MDOQ scores improved

Graham MD, Kemink JL, *Laryngoscope*, 1984
Welling DB, Nagaraja HN, *Otolaryngol Head Neck Surg*, 2000
Kato BM et al., *Otol Neurotol*, 2004
Sood AJ, Lambert PR, Nguyen SA, Meyer TA, *Otol Neurotol*, 2014

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ENDOLYMPHATIC SAC SURGERY

	DECOMPRESSION ODDS RATIO (95% CI) mean follow-up (range) n articles / N patients	SHUNT ODDS RATIO (95% CI) mean follow-up (range) n articles / N patients
COMPLETE OR SUBSTANTIAL VERTIGO CONTROL	79.3% (62.9% - 91.9%) 26.2mo (12-55mo) 6 / 267	76.4% (69.5% - 82.7%) 31.0mo (12-72mo) 19 / 1384
HEARING STABLE OR IMPROVED	72.8% (62.5% - 81.9%) 25.8mo (12-55mo) 8 / 303	71.4% (64.9% - 77.5%) 34.8mo (12-132mo) 14 / 799

Graham MD, Kemink JL, *Laryngoscope*, 1984
Sood AJ, Lambert PR, Nguyen SA, Meyer TA, *Otol Neurotol*, 2014
Bojrab DI 2nd, LaRouere MJ, Bojrab DI, et al., *Otol Neurotol*, 2018

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ENDOLYMPHATIC SAC SURGERY

	SHUNT WITHOUT SILASTIC ODDS RATIO (95% CI) mean follow-up (range) n articles / N patients	SHUNT **WITH SILASTIC** ODDS RATIO (95% CI) mean follow-up (range) n articles / N patients
COMPLETE OR SUBSTANTIAL VERTIGO CONTROL	75.0% (58.6% - 88.5%) 31.1mo (12-72mo) 5 / 467	76.9% (69.1% - 83.9%) 28.3mo (12-51mo) 14 / 917
HEARING STABLE OR IMPROVED	72.5% (59.4% - 84.9%) 32.6mo (12-72mo) 3 / 372	68.0% (62.7% - 72.1%) 32.5mo (12-132mo) 11 / 427

Sood AJ, Lambert PR, Nguyen SA, Meyer TA, *Otol Neurotol*, 2014

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ENDOLYMPHATIC SAC SURGERY

- EFFECTIVENESS: Decompression ≥ Shunt
- INTRALESIONAL STEROIDS: Steroids = No steroids
- HEARING: Decompression ~ Shunt
- HEARING: Shunt without Silastic > Shut with Silastic

Graham MD, Kemink JL, *Laryngoscope*, 1984
Sood AJ, Lambert PR, Nguyen SA, Meyer TA, *Otol Neurotol*, 2014
Bojrab DI 2nd, LaRouere MJ, Bojrab DI, et al., *Otol Neurotol*, 2018

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ENDOLYMPHATIC SAC VEIN DECOMPRESSION

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ELSVD

Effectiveness is heavily dependent on surgical technique

- A complete decompression is necessary for effective treatment
- ANATOMICAL LANDMARKS:
  - Donaldson's Line
  - Sigmoid Sinus
  - Jugular Bulb
  - Otic Capsule

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ENDOLYMPHATIC SAC VEIN DECOMPRESSION

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ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO

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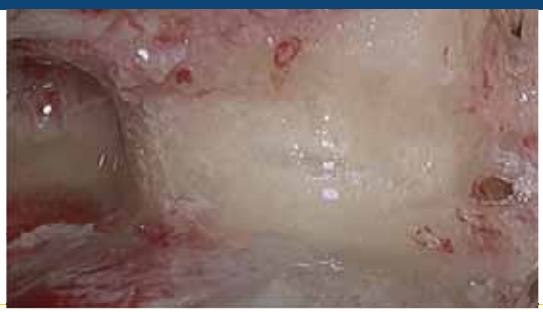
ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO



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ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO

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ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO

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ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO

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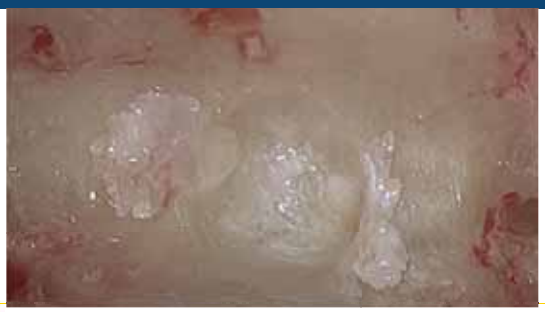
ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO



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ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO

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ENDOLYMPHATIC SAC VEIN DECOMPRESSION / PSCO

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THIRD ECHELON TREATMENTS

- VESTIBULAR NEURECTOMY
- LABYRINTHECTOMY

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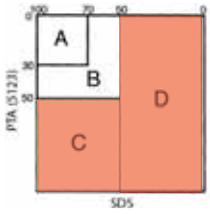
Criteria for Destructive Surgery

- 50/50 Rule



AAO-HNS C.H.E. GUIDELINES FOR HEARING PRESERVATION * *		
Class	4 Tone Average (dB HL)	Speech Discrimination (%)
A	≤ 30	≥ 70
B	30 – 50	≥ 50
C	> 50	≥ 50
D	any level	< 50

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What about bilateral disease?

- RULE: NEVER PERFORM DESTRUCTIVE SURGERY ON BILATERAL MENIERE'S PATIENTS
- Bilateral disease: up to 15-50% reported
  - most studies: 15-25%
- Latent contralateral ear manifests disease (HL, Sx) 90% within 5 years of ipsilateral ear
- Latent contralateral ear HL remains less severe than ipsilateral ear HL 90% of the time

Palaskas CW, Dobie RA, Snyder JM, *Laryngoscope*, 1988

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Vestibular Neurectomy

- Debate over relative effectiveness over labyrinthectomy
- Benefit over labyrinthectomy: preservation of hearing
- Risk of SNHL:
  - (highly) debated: from 0-25% reported
  - 10-25% of those occur immediately post-op
- Cons: craniotomy, CSF leak, brain retraction, chronic HA

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Vestibular Neurectomy

- Effectiveness: 80-90% (CHE class A/B/C)
- Non-responders:
  - inadequate resection of vestibular nerve
  - bilateral disease
  - misdiagnosis
- Cochleovestibular Nerve Section
  - no advantage over labyrinthectomy
  - persistent risks of craniotomy

Schmerber S et al., *Auris Nasus Larynx*, 2009

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Labyrinthectomy

GOLD STANDARD TREATMENT FOR MENIERE'S DISEASE

- "If you remove the organ, there can be no disease"
- Standard technique: blue-line the canals
- Modifications: Malcolm's Cup
- Complete extirpation of ALL FIVE neural elements is critical for effectiveness of procedure
  - 3 ampullae **cristae ampullaris**
  - spherical recess **saccule**
  - elliptical recess **utricle**

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Labyrinthectomy

- Cochrane Database:
  - Surgery for Meniere's Disease - 2013
  - "No prospective double blind RCTs"
    - Do you need a RCT for evaluate effectiveness of parachutes?

Pullens B et al., *Cochrane Database*, 2013

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Labyrinthectomy

- Effectiveness: 95+% (CHE class A/B/C)
  - degree of effectiveness is understated when reporting
  - ALL labyrinthectomy patients are class A - complete resolution
  - non-responders:
    - did not have adequate resection of neuroepithelium
    - may have bilateral disease
    - may be misdiagnosed

Kemink JL et al., *Otolaryngol Head Neck Surg*, 1989
Diaz RC et al., *Otol Neurotol*, 2007

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Labyrinthectomy

- Advantages:
  - extracranial, no craniotomy risks
- Cons:
  - complete hearing loss

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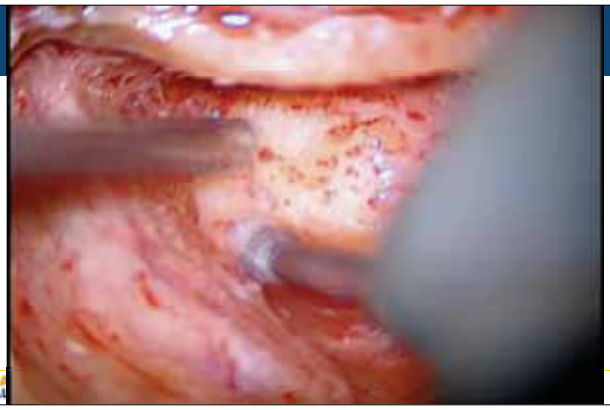

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QUESTIONS

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