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Endolymphatic Sac Tumors: An Overview

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Disclosure

- No relevant financial interests or other relevant relationships to disclose
- Will discuss off-label use of cochlear implants for single-sided SNHL

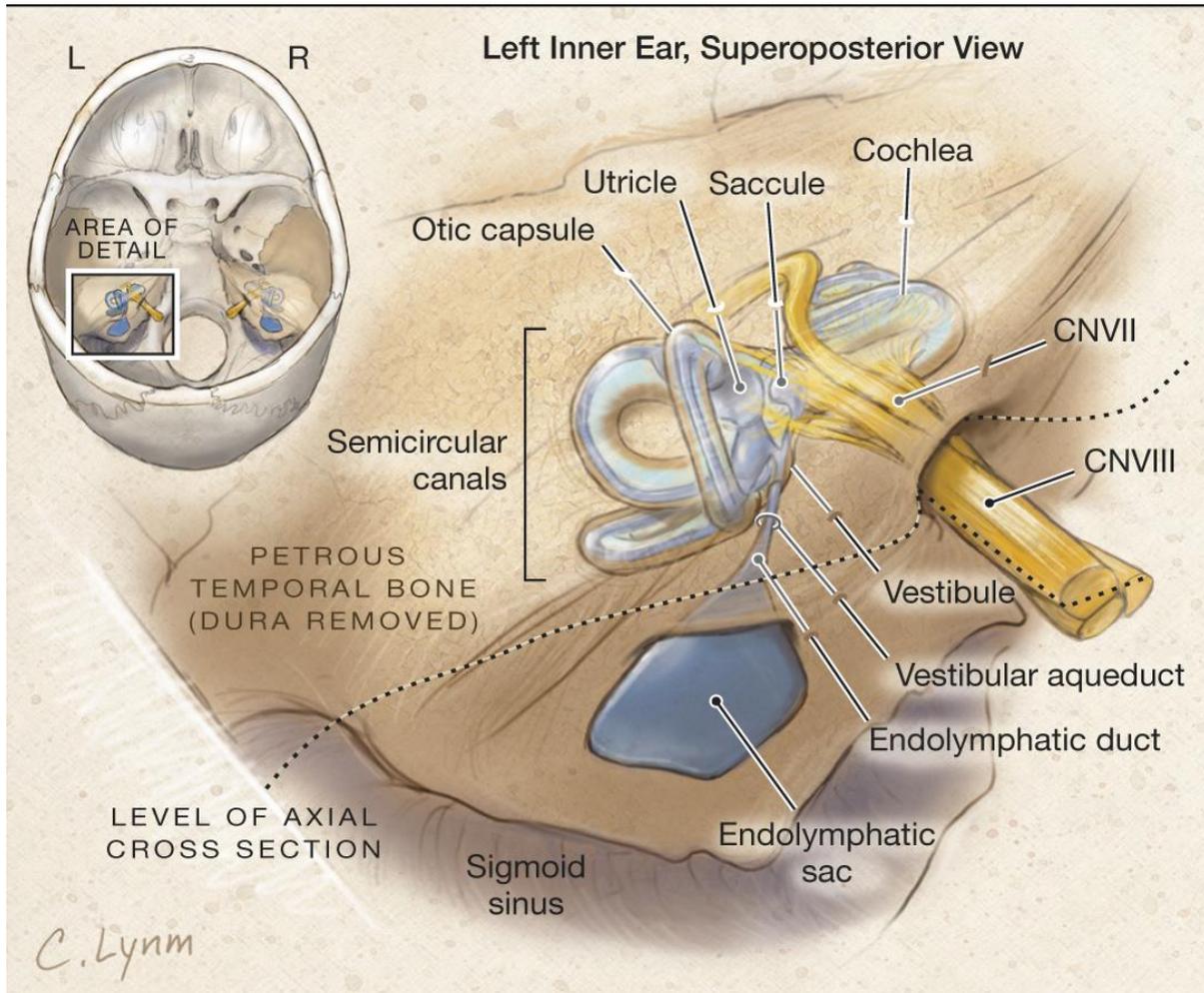


Outline

- What is an endolymphatic sac tumor?
- What are the symptoms?
- What is the work-up and treatment?



Endolymphatic Sac Anatomy



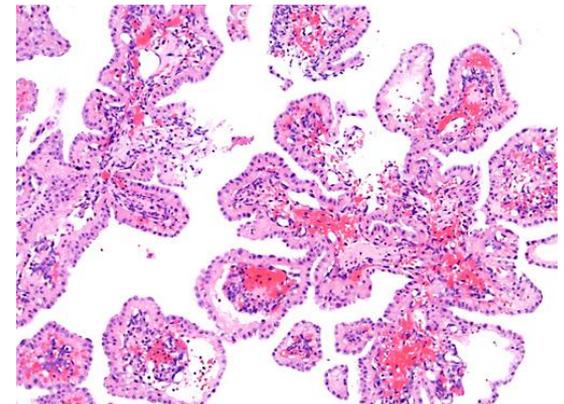
Endolymphatic Sac Function

- Part of the “membranous labyrinth”
- Filled with fluid called endolymph
- Secretes locally acting chemical called “saccin”
- Involved with inner ear fluid homeostasis, mechanisms not fully understood
 - ELS is involved with Meniere’s disease



ELS Tumors

- Extremely rare tumor originating from the endolymphatic sac, only recognized as a unique entity since 1989
- Benign (not cancer)
- Highly destructive, slowly progressive
 - Destroy bone of inner ear, around cranial vault/skull base, and around facial nerve
 - Can grow into nerves, pass through dura (sac around brain) and press on cerebellum
- May be asymptomatic until inner ear is partially destroyed
- Key: if hearing loss is present: high chance dura is invaded



Symptoms

- Most common presenting symptoms:
 - Hearing loss (85%)
 - Visible mass in the ear (50%)
 - Ringing in the ear (48%)
 - Facial paralysis (44%)
 - Dizziness/imbalance (44%)
 - Headache (37%)
 - Other neurologic (cranial nerve) weakness (25%)
- If present in both ears, outlook for balance is much worse



ELS Tumors and VHL

- 10-15% of VHL patients will have an ELS tumor
- Most of these are only one ear, but ~20% of VHL patients with an ELS tumor will have tumors in both ears
- Usually presents with hearing loss in 2nd, 3rd, or 4th decade of life
 - If not present by age 45, high unlikely
- Screening:
 - Hearing test at diagnosis
 - If abnormal, needs radiologic imaging with contrasted MRI and possibly CT
 - If normal, needs to be repeated at every 2-3 year intervals for surveillance until age 45
 - Past normal MRI or normal audiogram does not “clear” VHL patient from possibly having ELS tumor if patient is still relatively young



MRI of ELS



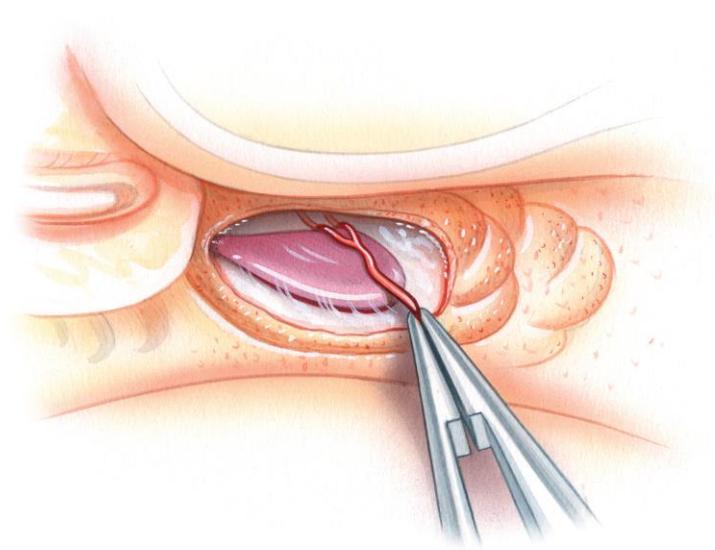
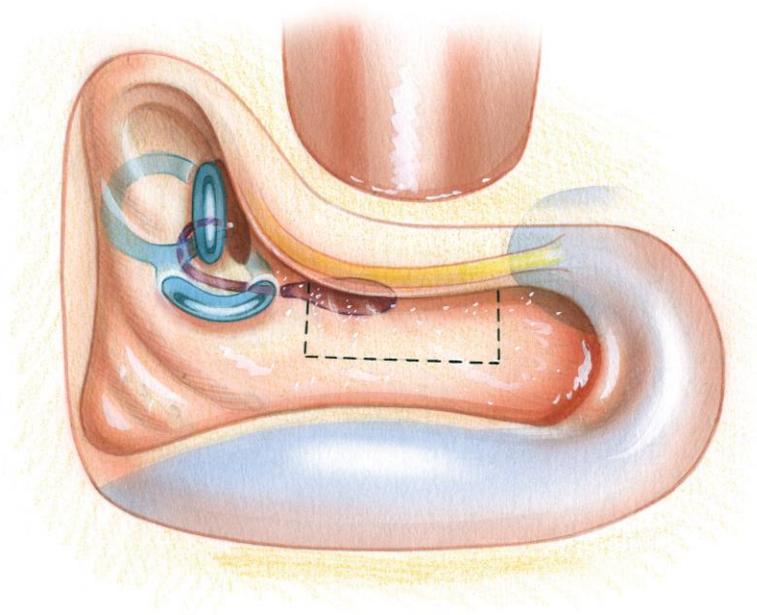
Treatment: Surgery

- Surgery is the primary treatment in most cases
- For very small tumors (rare):
 - Transtemporal (ear) approach, hearing preservation is possible
 - Removal of tumor and EL Sac (no tumor extension beyond)
 - Outpatient surgery, low risk, rapid recovery
- For most tumors:
 - Transtemporal (ear) approach +/- craniotomy
 - Hearing preservation not likely as part of the inner ear is commonly sacrificed
 - Tumor is removed with adjacent dura and dural repair
 - 2-5 day hospitalization, may have post-op imbalance, may need physical therapy
 - CSF leak possible
 - If facial nerve is involved with tumor, repair with nerve graft at same time



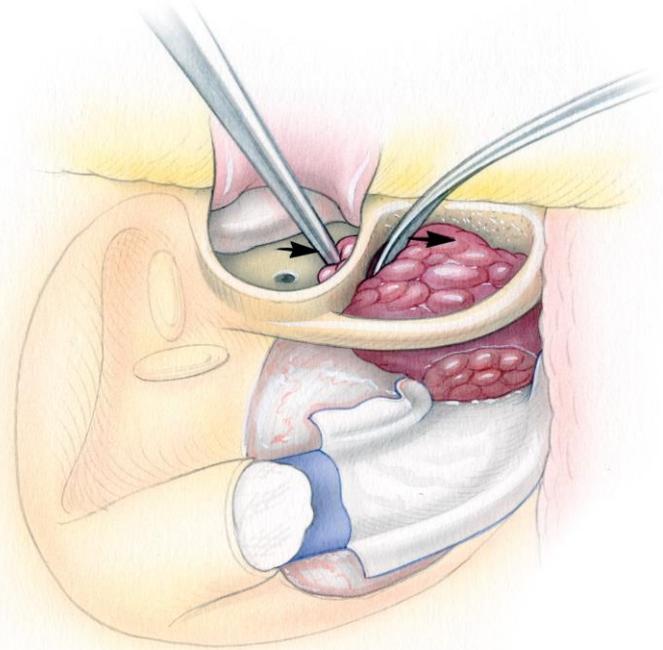
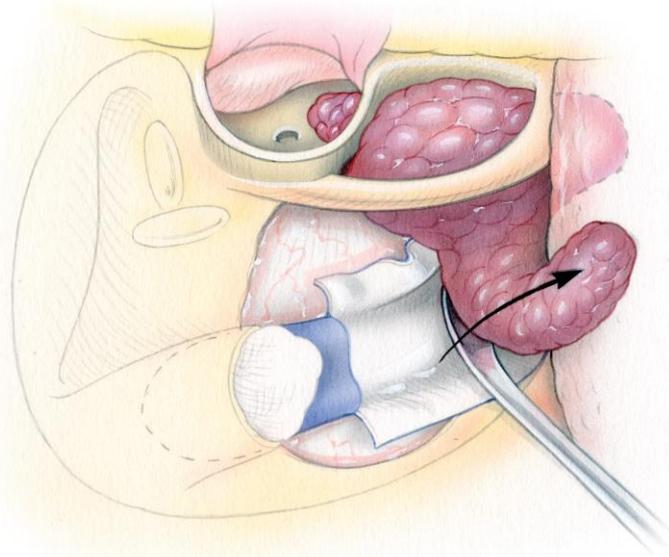
Transtemporal Approach (Mastoidectomy)

Images Courtesy of R. Jackler, Stanford U



Transtemporal Approach with Craniotomy

Images Courtesy of R. Jackler, Stanford U



Treatment: Surgery

- Early surgery is preferred:
 - Less tumor destruction
 - Less extensive surgery
 - Possibly lower recurrence
 - Prevent facial nerve involvement
- Recurrence rate after surgery fairly high (10-40%) depending on:
 - Size of the tumor
 - Extent of the tumor
 - Aggressiveness of surgery
- ***Biggest issue with failed surgery:*** surgeon not aggressive enough because they are dealing with a “benign” tumor; usually need to resect the adjacent dura (sac around brain) to completely remove the tumor (typically involves neurosurgeon)
- Surgery: ENT ear subspecialist (neurotologist) and neurosurgeon



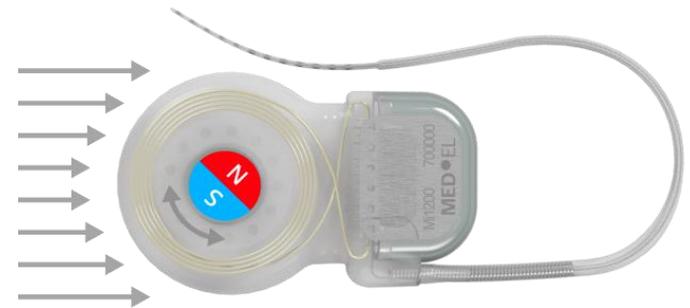
Chemo or Radiation Therapy for ELS

- Controversial, not enough data (mainly limited case reports)
- May be used as palliative treatment to control tumor (add-on after surgery), but not commonly accepted as primary treatment
- More likely to be used for a very large and or recurrent tumor



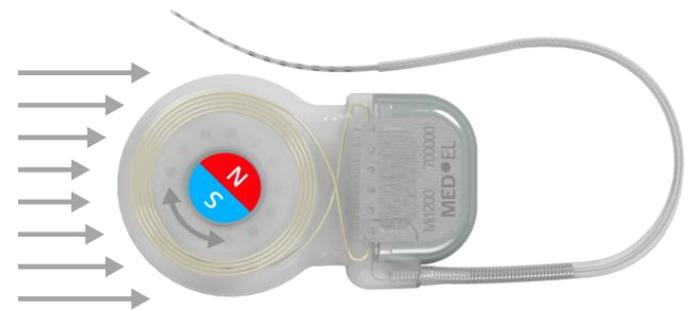
Cochlear Implants and VHL

- Obstacles:
 - MRI safety
 - MRI compatibility/image distortion
 - Risk/benefit in unilateral hearing loss



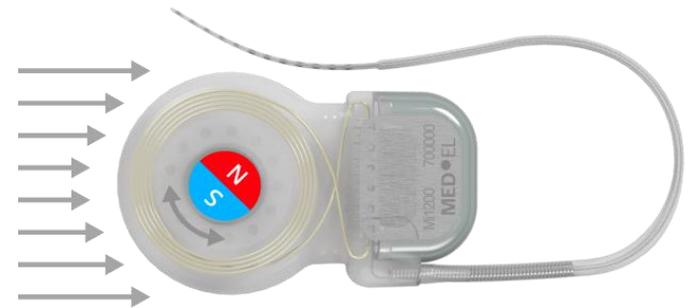
Cochlear Implants and VHL

- Obstacles:
 - MRI safety
 - Has internal magnet, risk of moving during MRI
 - Risk of depolarizing magnet
 - Earlier models not FDA approved with MRI unless internal magnet removed during separate minor surgical procedure and then replaced after imaging, but some attempts to wrap the head tightly and perform anyhow with mixed results
 - Current CI options now include compatibility without magnet removal for standard MRI at 3 Tesla (Med-El Synchrony)

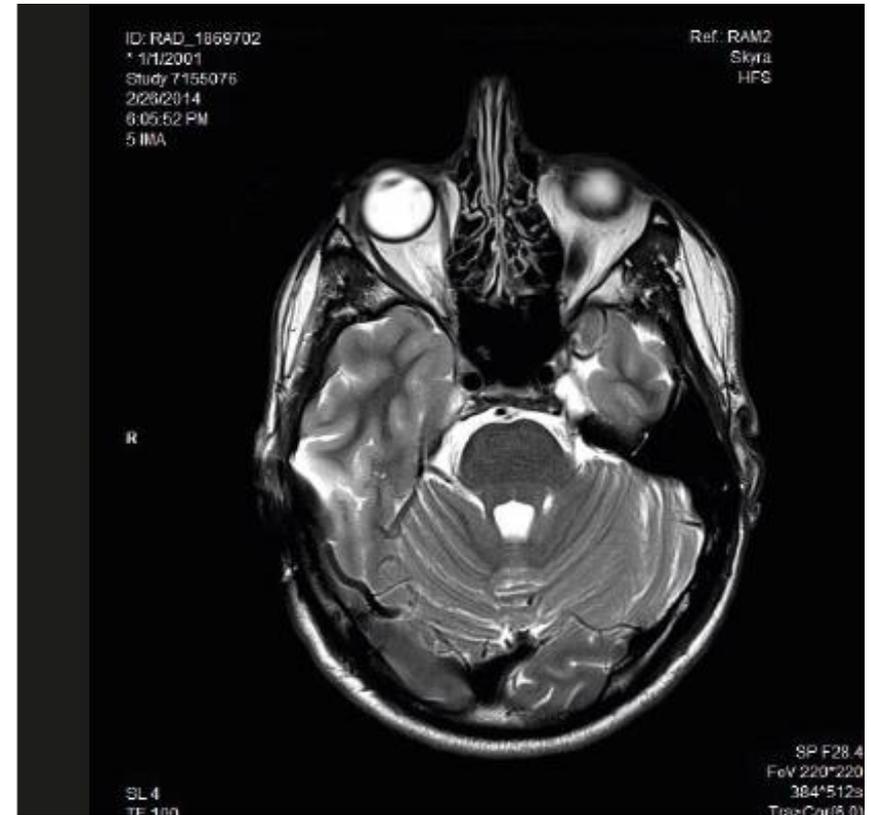
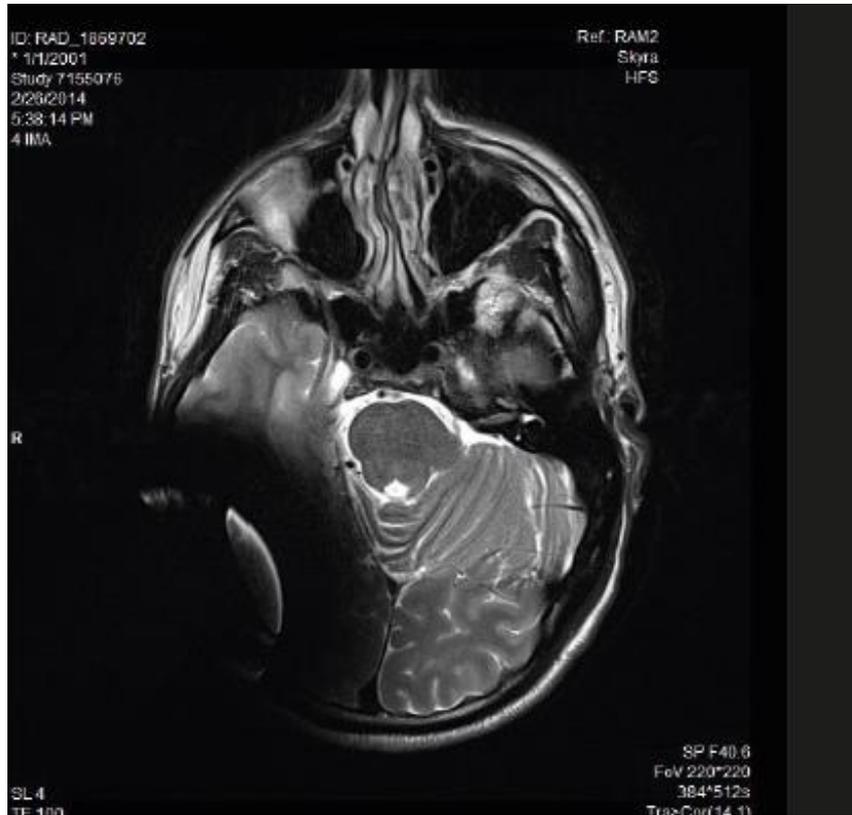


Cochlear Implants and VHL

- Obstacles:
 - MRI compatibility/image distortion
 - For traditional CI: If magnet not removed and MRI completed with head wrapped, major field distortion such that often more of half of the head not visible on MRI image; even if magnet was removed, still very prominent distortion
 - For current MRI compatible version, distortion is less than prior but still present when magnet not removed, but minimal when removed



MRI with Med-EI Synchrony (Magnet Removed Right)



Cochlear Implants and VHL

- Obstacles:
 - Risk/benefit in unilateral hearing loss
 - Traditionally, CI sound quality was only good enough that implantation was only recommended for bilateral complete or near-complete hearing loss
 - Technology/design/technique improved now such that CI for single-sided hearing loss is emerging; natural sound in one ear and “electronic” sound in another can merge well
 - Especially important when opposite ear may be at risk later in life; VHL patients at risk for bilateral HL
 - May need special insurance approval for unilateral hearing loss since CI is considered off-label in these patients



Cochlear Implants and VHL

- VHL patients may be considered for CI after counseling with multidisciplinary team based on:
 - Hearing status
 - Age
 - Disease manifestations
 - Projected frequency of future MRI
 - Underlying health
- It is possible to remove ELS tumor and place a cochlear implant at same setting, but may cause problems with surveillance imaging
- Alternate approach if the inner ear is involved is to insert a “place holder” within the inner ear during tumor resection to prevent future obliteration with scar/bone formation, then return later to place CI once confident that tumor is clear
- The sooner CI is applied/used after hearing loss, the better the hearing outcome



Conclusions

- Screen for ELS in patients with VHL with audiogram every 2-3 years until age 45
- Usually cause hearing loss as presenting symptom
- At time of diagnosis via hearing loss, high chance of dura involvement
- Treatment is usually surgery, which should be adequately aggressive due to potential for tumor recurrence
- Emerging role for cochlear implantation in management of ELS tumors in VHL

