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# Grand Rounds: Complex Hearing Aid Cases

Erica Bennett, Au.D, PhD

Chelsea Conrad, AuD

Brad Stach, PhD

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## Learning Objectives

As a result of this course, participants will be able to:

- Discuss amplification options for patients with unique audiological needs from complex medical diagnoses including semicircular canal dehiscence, usher syndrome, MELAS, etc.
- Discuss CROS/BiCROS options for patients with traditional hearing aids as well as patients with cochlear implants.
- Explain benefits of traditional amplification versus implantable devices and when to consider surgical intervention.

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## Amplification Options for a Patient with Usher Syndrome

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## Usher Syndrome

- Type I:
  - Typically born profoundly deaf
  - Usually present with balance disorders
  - Vision loss in the first few years of life
- Type II
  - Typically born with lesser hearing losses (moderate-severe)
  - Vision loss develops more slowly
  - Normal balance
- Type III
  - Vision and hearing loss develop later in life (usually beginning late childhood, early adulthood)
  - May also develop balance disorders

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## Ms. W

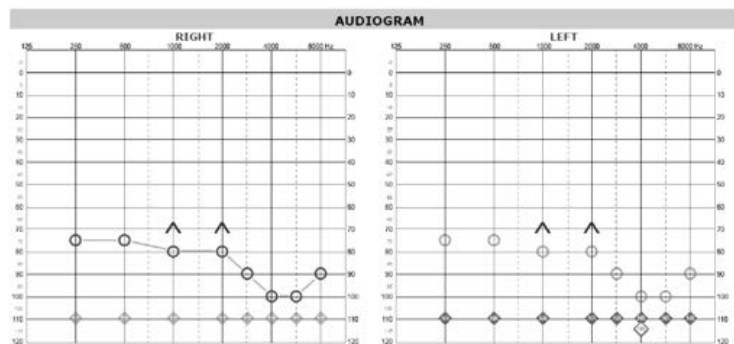
- 64 year old female
- Presented to clinic with concerns of progressive hearing loss and balance issues
  - Patient also reported:
    - Constant bilateral tinnitus
    - Dizziness
- History of progressive vision loss
- Former heroin addict

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## Ms. W

- Otoscopy: normal bilaterally
- Immittance: Type A tympanogram with absent reflexes bilaterally
- Word recognition scores: 28% right ear and 0% left ear
- Absent DPOAEs bilaterally



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## Ms. W

- Recommendations:
  - Consider CI evaluation
  - Balance Function Test
  - Hearing Aid Selection

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## Hearing Aid Selection

- Discussed cochlear implants with the patient and her niece in great detail
- Patient was opposed to surgery at this time
- Super power BTE with earmold was chosen for right ear only

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## Hearing Aid Delivery

- Patient was fit with BTE for right ear only
- Reported comfortable fit and improved hearing
- Patient was scheduled to return for a two week check, but did not return for 2 years
- 2 years later...
  - Patient's vision had significantly declined
  - Diagnosed with Usher Syndrome
  - Hearing aid was not working
  - Patient was interested in CI

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## Cochlear Implant Evaluation

- Testing was completed with clinic's power BTE hearing aids and comply tips
- Aided Thresholds

	250	500	1000	2000	3000	4000	6000
Right	NR	60	45	40	55	60	NR
Left	NR	NR	NR	NR	NR	NR	NR

- Speech Perception Results

Test	Right Ear (best aided)	Left Ear
HINT in Quiet	32%	0%
AZ Bio in Quiet	5%	0%
CNC in Quiet	22%	5%

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## Cochlear Implant Discussion

- Discussed all three manufacturers
  - Advanced Bionics Neptune
  - Remote microphone
  - ComPilot
  - TV Link

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## Surgical Consultation

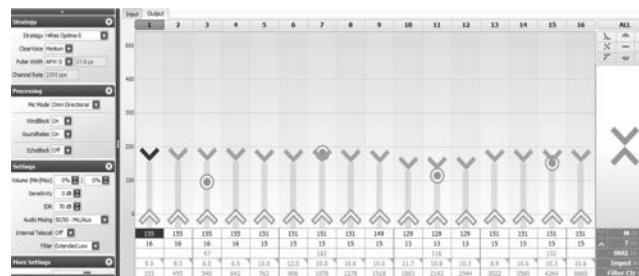
- At initial surgical consultation, patient presented with elevated Creatine
- Surgeon concerned with renal failure
  - Patient referred to Nephrologist/primary care provider for clearance
- Patient was seen two months later with clearance; however, patient was hypertensive and surgery could not be scheduled until blood pressure was maintained
- Finally, 6 months following initial CI evaluation, Ms. W had surgery

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## Ms. W Initial Activation

- NRI revealed all electrodes were functioning
- Patient was not able to loudness scale; therefore live speech/speech bursts were used to program
  - Patient was provided with progressive MAPs
- Patient had very thick hair; therefore, 4 internal magnets with 1 external magnet were required



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## Ms. W Follow-up Appointments

- Patient seen at 1 week, 1 month, 3 months, and 6 months
  - Programming using speech bursts was completed at each visit
  - Aided audiogram demonstrated responses between 25-35 dB HL from 250-4000 Hz
  - Ms. W mentioned she was considering shaving her head
  - Recommend she come in for magnet evaluation the day she changes her hair style
- At 6 month visit speech perception tests were completed:

Test	Implant alone
HINT in Quiet	92%
AZ Bio in Quiet	56%
CNC in Quiet	55%

- Discussed option of hearing aid for contralateral ear and/or bilateral implants

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## Ms. W Explant/Reimplant

- Patient did not return for several months
- She finally returned due to pain at the magnet site
- She shaved her hair and had not returned for evaluation of magnet strength
- She was seen by ENT and they prescribed antibiotics, instructed patient NOT to wear implant
- Patient continued to wear the implant and an explant/reimplant was necessary

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## 2<sup>nd</sup> Device Discussion

- Advanced Bionics Naida Q90
- Options for contralateral ear:
  - Naida Link Ultra Power
  - Naida Link RIC
  - Naida Link CROS
  - Eventually bilateral implant
- Hearing evaluation showed no measureable responses bilaterally

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## Ms. W 2nd Activation

- Patient was able to understand words and distinguish voices at the activation appointment.
- Patient stated she was planning to shave her head again
  - Scheduled a magnet check appointment directly following hair appointment

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## Ms W. Follow-Up: 3 weeks

- Magnet strength was checked following hair change and was deemed to be appropriate
  - Patient decided she will keep head continuously shaved
- Patient fit with CROS device
- Emerging speech understanding assessed:
  - Ling Sounds: 6/6
  - Closed Set Spondees: 6/6

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## Ms. W: Follow-Up Phone Call

- Patient's niece reports CROS device is a great improvement, particularly when sitting around the dinner table
- Patient is doing well with implant and CROS device and will return soon for 3 month follow-up.

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## Take Away

- Multiply involved patients require creative solutions
- The CROS device is a good contralateral option for patients who cannot receive surgery or do not wish to have a second surgery.

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## Wrap Up

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Amplification options for a  
patient with MELAS

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## MELAS: Mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes

- Caused by mutations in the genes in mitochondrial DNA
  - Patients inherit altered gene from mother
- Most patients have a build up of lactic acid in their bodies, which can lead to:
  - Fatigue
  - Vomiting
  - Difficulty communicating (aphasia)
  - Muscle weakness
- Patient also experience stroke-like episodes
- Hearing loss is common in these patients although the etiology is unclear

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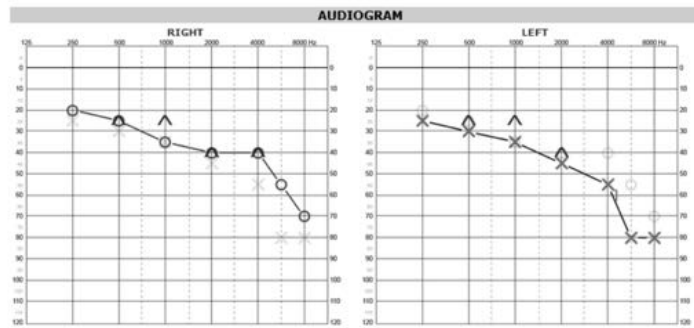
## Ms. F

- 29 year old female
- Presented with concerns of hearing loss
  - Denied any other auditory symptoms

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Ms. F

- Otoscopy: normal bilaterally
- Immittance: normal middle ear function bilaterally
- Word recognition presented at 80 dB HL
  - Excellent bilaterally



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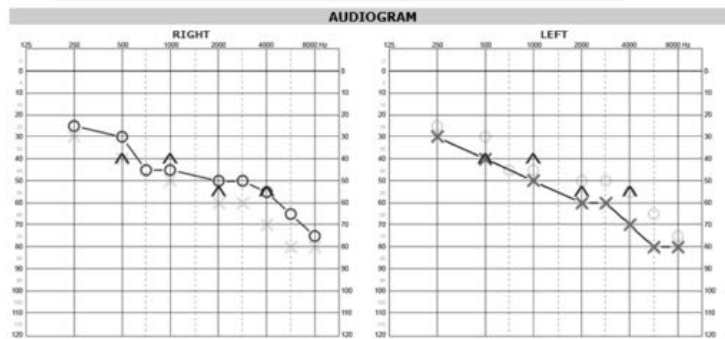
Ms. F Recommendations

- ENT for evaluation
- Further evaluation of asymmetry if indicated
- Recommend patient schedule hearing aid selection appointment pending medical clearance

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## Ms. F: 8 years later...

- Ms. F is 37 years old
- Ms. F is in school for nursing and working as a CNA
- Word recognition presented at 80 dB HL is excellent bilaterally



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## Ms. F Recommendations

- ENT for evaluation
- Further evaluation of asymmetric hearing loss and absent reflexes
- Recommend patient pursue hearing aids through Michigan Rehab Services (MRS)
- ENT recommended an MRI but patient declined at this point in time and would rather take an observation approach

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## Ms. F Hearing Aid Selection

- Patient selected bilateral open fit BTE hearing aids
- An FM system for classroom use was also requested for the patient through MRS

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## Ms. F Hearing Aid Delivery

- Patient reported hearing aids were loud, but clear
- Probe tube measurements were consistent with results expected from the hearing aids
- Practiced use of FM system in office
- Patient encouraged to return for a check in two weeks

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## One year later...

- Ms. F returns with concerns of a non-functioning FM system
- She reports she has not been using the FM system at school
  - Lengthy discussion of advocating for self
  - Counseling and practice to increase comfort with FM system
- Recommended an updated hearing test and follow-up hearing aid check
  - Patient was uninsured but test could be completed through MRS

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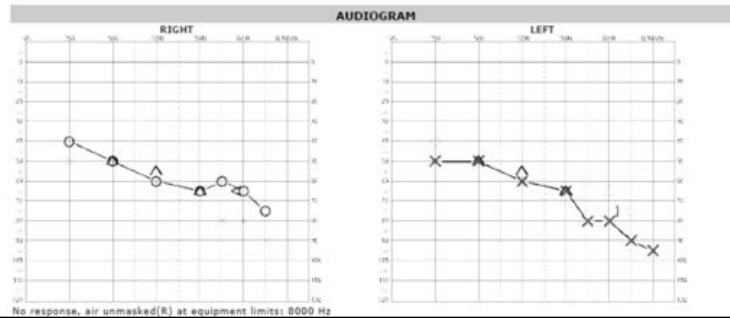
## 6 years later...

- Ms. F returns to the clinic, accompanied by her mother, with reports of decreased hearing following a stroke
- The patient has also been diagnosed with Wernicke's Aphasia following the stroke
  - Wernicke's aphasia (aka fluent aphasia): ability to grasp spoken words or sentences is not well maintained
- Patient's mother reports memory loss and confusion

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## Ms. F

- Otoscopy: normal bilaterally
- Immittance: type A tympanograms with absent reflexes bilaterally
- Word recognition attempted but could not be completed due to patient frustration
- Hearing had decreased significantly since previous evaluation



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## Mrs. F Recommendations

- Follow-up with ENT
- Patient's insurance stopped paying for speech and language therapy
  - Patient referred to Wayne State University stroke clinic
- Schedule hearing aid check
  - Patient's mother reported patient received other hearing aids from outside vendors. Patient instructed to bring in all hearing aids she owns

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## Ms. F Hearing Aid Check

- Patient brought in several hearing aids, but none were functioning
  - Patient is no longer eligible for services through MRS, as she is not working
- She was fit with donated BTEs with slimtubes and small power domes
- Patient was able to independently change batteries and insert hearing aids in her ears

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## 2 months later...

- Patient returned to clinic following several more stroke-like episodes with concerns of decreased hearing
  - Also presented with balance concerns
- Patient diagnosed with MELAS

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## Ms. F: Recommendations

- Hearing was unchanged. Therefore recommendations were:
  - Patient referred for balance function test
  - Patient had been regularly attending stroke clinic at Wayne State University
  - Recommended patient continue speech and language therapy

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## Ms. F: Balance Function Test

- Patient was given complete test battery including:
  - VNG
  - Rotational Chair
  - oVEMP and cVEMP
  - vHIT
  - Posturography
- All results were normal except Posturography
- Posturography revealed a severe sensory dysfunction pattern
- Patient referred to physical therapy for falls safety

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## Further hearing recommendations

- Would Ms. F be a CI candidate if hearing continues to decrease?
  - Is hearing loss retrocochlear or cochlear in origin?
- Research shows:
  - Hearing loss is likely cochlear in origin (Sue, Lipsett, Crimmins, 1998)
  - Several patients with MELAS have been implanted yielding good results (Karkos, Anari, & Johnson, 2005; Hill, Wintersgill, & Scott, 2001; Rosenthal, Kileny, Boerst, 1999)
  - However, research recommends acoustic MRI in addition to CT prior to implantation to assess integrity of auditory system (Karkos, Anari, & Johnson, 2005)

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## Take Away

- Once again, multiply involved patients require creative solutions
- Patients with MELAS may be candidates for cochlear implantation provided there are no known retrocochlear pathologies

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## Wrap Up

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## Hearing Aid Fitting with Superior Semicircular Canal Dehiscence (SSCD)

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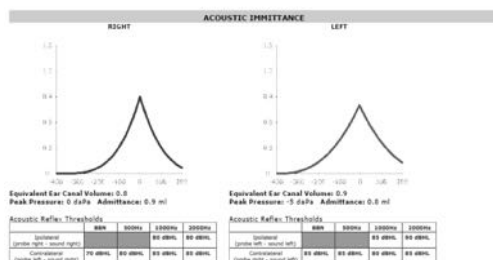
## Patient C

- 11 year old male, accompanied by his mother, with concerns of left ear hearing loss
  - Also reported sensitivity to loud sounds
- Patient's mother reported something was "malformed" in the child's left ear but could not describe further
- Wears a hearing aid in the left ear, reported as uncomfortably loud
- Interested in a new hearing aid prior to start of the school year

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## Patient C

- Otoscopy: clear ear canals bilaterally
- Immittance: normal middle ear function bilaterally



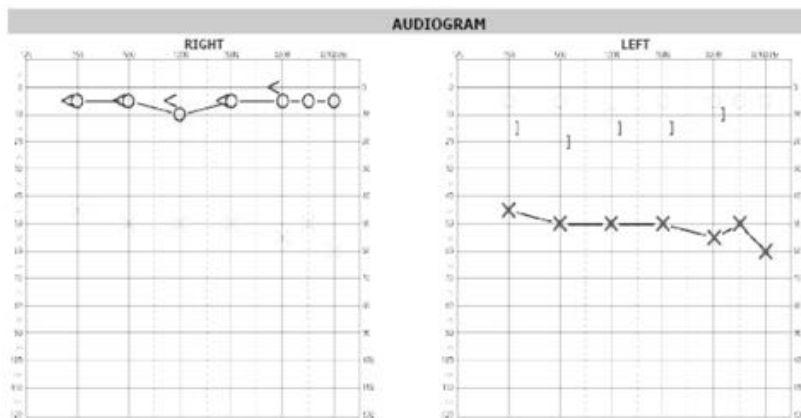
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## Patient C

- Excellent word recognition bilaterally
- DPOAEs: present right ear and absent left ear



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continued



## Patient C: Recommendations

- Continue to appointment with ENT
- Further testing for the left ear if indicated
  - Consider VEMP testing
- Pending medical clearance, schedule hearing aid selection and/or evaluation of current hearing aid.
- Request outside records as patient's mother reported patient had a CT scan.

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## Outside CT scan

- Bilateral SSCD
- Left ear Mondini malformation

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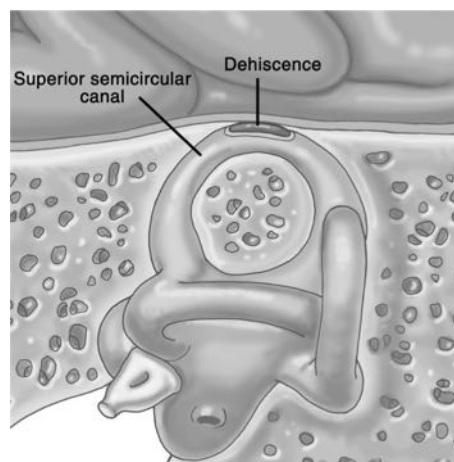
## Mondini Dysplasia

- 1.5 turns of cochlea rather than 2.5
- Hearing loss typically presents as profound SNHL, but can cause hearing loss of varying degrees

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## Superior Semicircular Canal Dehiscence (SSCD)

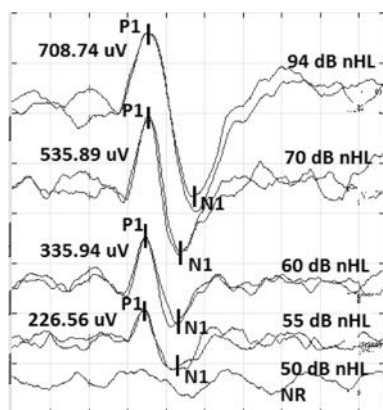
- Dehiscence in the bone covering semicircular canal
- Auditory Symptoms:
  - Conductive hyperacusis (increased sensitivity to bone conducted sounds)
  - Tinnitus
  - Autophony
- Vestibular Symptoms:
  - Tullio phenomenon: vertigo in response to loud sounds
  - Unsteadiness



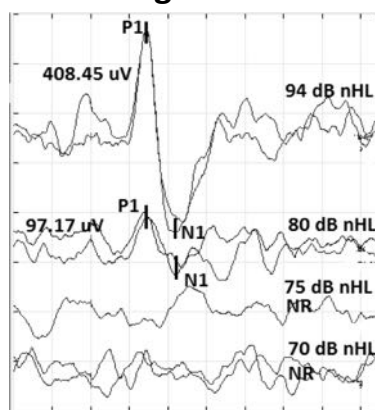
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## Patient C: VEMP Testing

### Left Ear



### Right Ear



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## Hearing Aid Fitting

- Patient has a hearing loss in the left ear that is likely sensorineural due to Mondini malformation
- However, hearing test results make the hearing loss appear to be conductive due to enhanced bone responses from the dehiscence
- How should this patient be fit?

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## Evaluation of current hearing aids

- Patient's current hearing aid was assessed
- Patient's hearing aid was set as if it were a conductive hearing loss, thus providing the patient with excessive amplification in an already sensitive ear
- Patient's current hearing aid was re-programmed without the bone conduction thresholds and the patient reported increased comfort with the sound
- Patient's hearing aid met DSL targets for a sensorineural hearing loss

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## Take Away

- Patients with SSCD, who also have a hearing loss, should be amplified with caution, due to sensitivity to sound
- Patients with SSCD should not be treated as if they have a conductive hearing loss

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## Wrap Up

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## Amplification Options for Patient with an Acoustic Schwannoma

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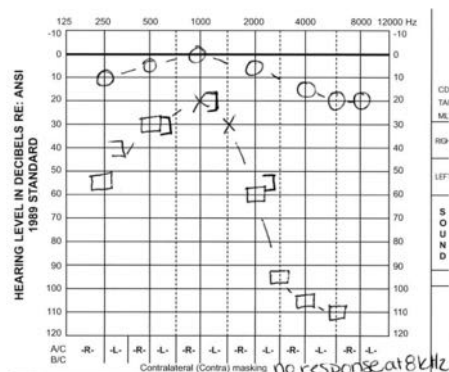
### Ms. M

- 37 year old female
- Came to clinic for a 2<sup>nd</sup> opinion
- Presents with left sided hearing loss, vertigo, and left sided tinnitus
- Brought with her a 2 year old hearing evaluation from an outside clinic

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## Ms. M: outside hearing evaluation

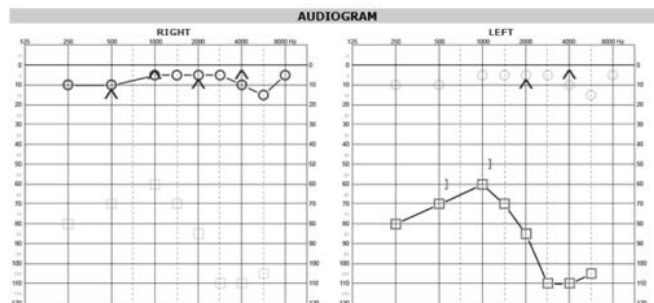
- Otoscopy: reported as normal bilaterally
- Immittance: not completed
- Word Recognition Testing:
  - Right and Left ear: 100% at 50 dB HL



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## Ms. M

- Otoscopy: revealed clear ear canals bilaterally
- Immittance:
  - Normal Type A tympanograms bilaterally
  - Present right ipsilateral reflexes, all other reflexes were absent
- Word Recognition Testing:
  - Right ear at 80 dB HL: 100%
  - Left ear at 90 dB HL: 54%
  - Left ear at 80 dB HL: 16%
- Negative Stenger



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## Ms. M Recommendations

- Follow-up with ENT as planned
- Further testing due to retrocochlear signs and symptoms
- Recommended Balance Function Test
  - Patient scheduled but did not attend
- Consider amplification needs pending medical clearance

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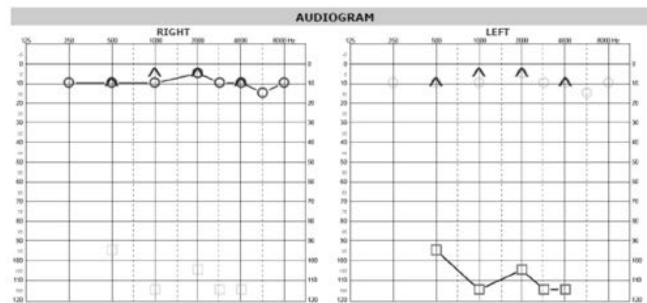
## Ms. M: Acoustic MRI

- Imaging revealed a 1.5 mm vestibular schwannoma on the left side
- Vestibular Schwannoma: slow growing tumor originating from the overproduction of Schwann cells
- Decided to take an observation approach
  - Recommended annual hearing/ENT evaluations or sooner if changes noted

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## Two years later...

- Patient returns due to decreased hearing in the left ear
- Recommended follow-up MRI, which was stable
- Patient to consider amplification options



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## Amplification options

- Traditional hearing aids or a cochlear implant are not recommended as patient has a retrocochlear pathology and hearing loss is not cochlear in origin
- Patient to consider:
  - CROS system
  - BAHA

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## Ms. M: BAHA Evaluation

- Testing completed using Az Bio Sentences in a variety of conditions
- Patient subjectively reported decreased listening effort and increased clarity with the bone oscillator
- Objective results:

Signal/noise (dB HL)	Without oscillator	With oscillator
55/40	100%	DNT
55/45	86%	100%
55/50	63%	79%
55/55	0%	11%

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## Ms. M: BAHA

- Patient chose Baha Connect
- Surgery completed without complications
- Patient fit with BAHA 12 weeks post surgery
  - Follow-up 1: Patient reported that device had changed her life and requested overall gain be increased slightly.
  - Follow-up 2: Patient scheduled but did not attend and has not been in clinic since initial follow-up

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## Take Away

- Traditional amplification and/or cochlear implants may not be appropriate for retrocochlear losses
- Baha can be a great option for patients with single sided deafness

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## Wrap Up

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## Amplification: BiCROS vs. Bilateral Hearing Aids

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### Mr. F

- 65 year old male
- Presented to clinic with concerns of hearing loss, dizziness, and tinnitus
- Patient is a musician and reported distortion of music

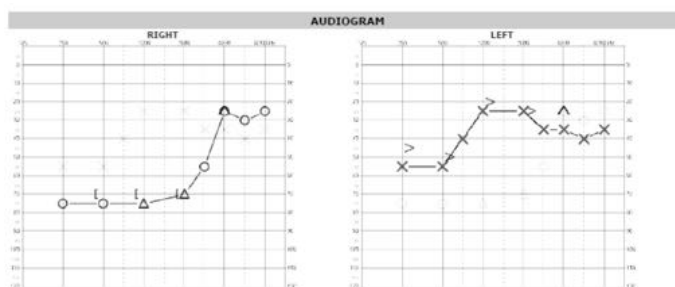
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Mr. F

- Otoscopy: normal bilaterally
- Immittance:
  - Right ear: type A tympanogram and absent reflexes
  - Left ear: normal middle ear function
- Word Recognition Scores:
  - Left ear: 100% at 80 dB HL
  - Right ear: 10% at 90 dB HL and 20% at 100 dB HL



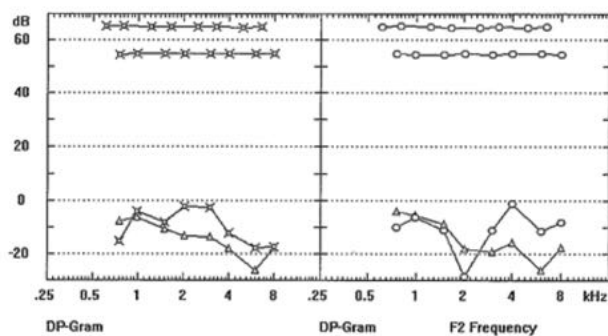
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Mr. F

- In addition, DPOAE testing was completed and results were consistent with pure tone audiogram.



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## Mr. F Recommendations

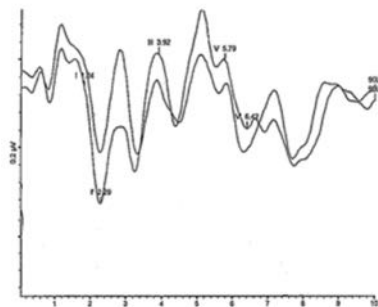
- Follow-up with ENT as planned
- Consider balance function test
- Consider amplification needs
- Further testing if clinically indicated due to retrocochlear signs and symptoms

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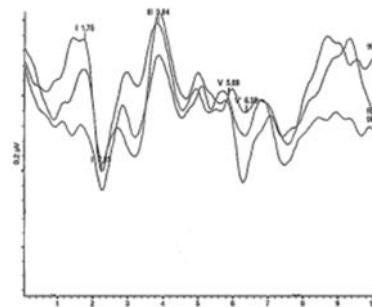
## Mr F: ABR

- Click Stimuli of rarefaction polarity, presented at a rate of 21.1 at 90 dB HL were used to obtain the following results:

**Left Ear**



**Right Ear**



- ABR shows well-formed responses to click stimuli at normal absolute latencies and interwave intervals. There was no electrophysiological evidence of VIIIIn or auditory brainstem pathway disorder.

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## Mr. F: Hearing Aid Selection

- Amplification options:
  - Bilateral Hearing Aids
  - BiCROS System
  - Left ear hearing aid only
- Options were discussed with the patient
- Counseled on “trial and error” approach to figuring out the correct option for his unique hearing loss
- Patient chose BiCROS system

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## Mr. F: Hearing Aid Delivery 1

- Patient was fit with a BiCROS system
- Right ear: CROS
- Left ear: Level 2 RIC with a closed dome
- Probe tube measurements were consistent with results expected from the hearing aid
- Patient reported good sound quality and was satisfied with devices.

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## Mr. F: Follow-Up 1-BiCROS

- Patient reported an echo sensation and dislike of his own voice
  - Dome changed from a closed to an open dome
  - Occlusion compensation set to strong per manufacturer algorithm
- In addition, he felt too much information was being sent from the CROS
  - CROS balance was changed to decrease volume of CROS device

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## Mr. F: 2<sup>nd</sup> Follow-up BiCROS

- Patient reported echo sensation was better
- He reported increased difficulty hearing in background noise, specifically at work
- He also requested to try the tinnitus feature in the left ear due to persistent tinnitus (mainly on right)
  - Patient counseled on use of tinnitus maskers, but was provided with a tinnitus program for the left ear
- Patient unsure if he liked the device; therefore trial period was extended.

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## Mr. F: 3<sup>rd</sup> Follow-Up BiCROS

- Patient wishes to upgrade technology to increase performance in background noise
- Patient upgraded to a level 3 device in the left ear
  - Realistic expectations, particularly in background noise, were discussed

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## Mr. F: 1<sup>st</sup> Follow-Up with Upgrade

- Patient reports background noise is still not better
- Patient still has complaints of tinnitus
- Patient also reports that he misses hearing out of two ears
- Bilateral amplification is ordered for patient
  - Patient chose bilateral RIC hearing aids.
    - Right ear: acrylic micromold with canal lock with power receiver
    - Left ear: medium open dome and standard receiver

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## Mr. F: Bilateral Hearing Aid Delivery

- Patient is extremely satisfied with sound quality
- Music was played in the office and patient reported better sound quality with bilateral amplification
- He was given volume control incase sounds were too loud in the real world

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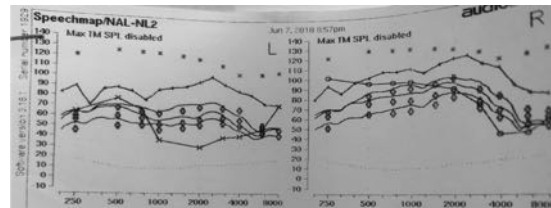
## Mr. F: Follow Up: Bilateral

- Patient reports significant improvement in noise, as well as resolution of tinnitus when wearing the devices

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## Mr. F: Follow-Up Bilateral

- Probe tube measurements completed



- Aided outcome measures:

Condition	Left Hearing Aid	Bilateral Hearing Aids
Quiet	99%	100%
+ 10 SNR	60%	62%

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## Take Away

- Sometimes you have to try MANY things before you get it right
- BiCROS is not always the best option for patients with asymmetric hearing loss
- Bilateral amplification may reduce tinnitus in the poorer ear for patients with asymmetric hearing loss
- Consider functional testing to assess patient performance as well as patient's preference

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## Wrap Up

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## Amplification for a Patient with Unrealistic Expectations

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continued



## Mr. K

- 82 years of age
- Long term hearing aid user
- Annual hearing evaluation
  - Concerns of decreased hearing
  - No other auditory concerns

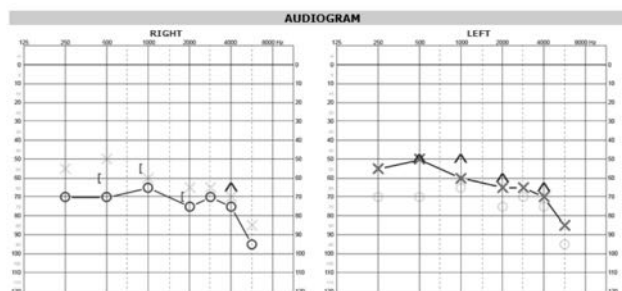
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## Mr. K: Hearing Evaluation

- Otoscopy: monomeric right TM, normal left
- Immittance:
  - Right ear: Type D tympanogram with absent reflexes
  - Left ear: Type A tympanogram with absent reflexes
- Word Recognition:
  - Left ear: 80% at 80 dB HL
  - Right ear: 8% at 80 dB HL
  - Right ear: 26% at 90 dB HL



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continued



## Mr. K Recommendations

- Follow up with ENT
- Consider further testing due to asymmetric word recognition scores
- Patient is unhappy with current hearing aids
  - Schedule hearing aid check
  - Consider new hearing aids

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## Mr. K Hearing Aid Check/Selection

- None of Mr. K's hearing aids were functioning
- New hearing aids chosen
  - Bilateral BTEs with full shell earmolds
  - Telecoil for phone
- Patient was extremely satisfied with new hearing aids
- Recommended annual checks or sooner if necessary

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continued



## Mr. K: 5 years later...

- Came in a handful of times for new earmolds, tubing changes, etc.
  - Did not schedule or attend regular hearing evaluations, although they were recommended
- Came to clinic for ENT/hearing evaluation reporting right ear pain
  - HT cancelled due to right otitis externa and significant drainage
  - Patient advised not to wear his hearing aid in the right ear

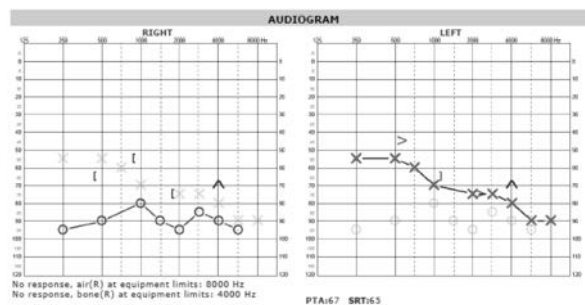
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## Mr. K: 1 year later...

- Came to clinic reporting right ear pain
- Otoscopy: perforation right ear and normal left ear
- Immittance:
  - Right ear: Large volume with absent reflexes
  - Left ear: Type A tympanogram with absent reflexes
- Word Recognition:
  - 0% in the right ear at 90 dB HL (patient refused higher levels)
  - 32% in the left ear at 90 dB HL



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continued



## Mr. K: Recommendations

- Follow up with ENT
- Retest post medical management of the right ear
- Consider cochlear implant evaluation
- Schedule Hearing Aid Selection

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## Otolaryngology Follow-Up

- Patient has 40% perforation of right TM
- CT scan revealed the following:
  - Right TM perforation with mild soft tissue density surrounding the middle ear ossicles, suggesting inflammatory debris
  - Superior semicircular canal dehiscence on the left
  - No acute intracranial abnormality seen
- Recommended surgical repair of perforation
  - Not for hearing purposes but for infection control

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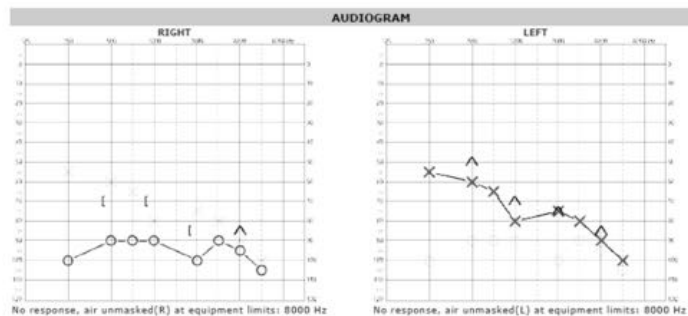
## Mr. K: Hearing Aid Selection

- Amplification Options:
  - Cochlear implant
  - Bilateral hearing aids
  - Hearing aid left ear only
- Patient fit with a left super power BTE with full shell earmold

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## Mr. K: 4 years later...

- Returns to clinic with left ear pain, this time pain is located on helix of left pinna
- ENT orders biopsy
- Hearing is unchanged



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## Mr. K Biopsy

- Biopsy revealed: basal cell carcinoma on L antihelix
- Patient declined surgery as it would require him not to wear his left hearing aid until the ear healed
- Drainage issues have resolved in the right ear
- Patient is now interested in new hearing aids and is medically cleared by ENT

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## Mr. K Hearing Aid Selection

- Patient came to clinic with brochure regarding Bluetooth technology, as his friend had great success with Bluetooth compatible hearing aids.
  - He was only interested in RIC hearing aids with connectivity to the iPhone
  - Patient did not have an iPhone
  - However, his friend stated they “changed his life”
- Difficult discussion regarding success with hearing aids and what hearing aids were appropriate for his hearing loss
  - Also discussed the limits of what traditional amplification could provide
- Amplification options
  - Cochlear Implant
  - Bilateral hearing aids
  - Hearing aid left ear only

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## Mr.K

- Would not fit patient with bilateral RIC hearing aids, as they were not appropriate for his hearing loss and/or chronic drainage issues
- Patient chose bilateral super power BTE hearing aids with full-shell earmold
- Ordered streamer for his Android cellphone

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## Mr. K Follow-Up

- Patient reported he is hearing much better with the hearing aids
- Subjectively, patient can have a conversation in office without written support
- Patient was still having difficulty on the phone
  - Ordered patient a CapTel phone (he had declined up until this point)

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## Mr. K Phone Options

- Patient chose a CapTel phone with Bluetooth technology
- This allowed the patient to stream his phone calls with the assistance of visual aid
- Patient reported improved listening effort on the phone
  - Allows him to converse with his many grandchildren around the country

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## Take Away

- As we know, sometimes what the patient wants is not always best
- Sometimes we have to come up with ways to compromise in order to meet patient's needs
- Counseling is extremely important when patient has unrealistic expectations
- Bluetooth CapTel phones can be used with Bluetooth capable streamers, which can aid patient on the telephone

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continued



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# Wrap Up

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