Advanced Diagnostic Audiometric Testing to Aid in Differential Diagnosis, Measure Patient Outcomes, and Interesting Case Studies

Abstract:
This presentation will focus on advanced audiometric techniques that will aid in differential diagnosis of multiple audiological disorders. We will discuss advanced immittance testing, advanced audiometric testing, and advanced real ear measures and techniques for difficult to solve hearing aid cases and programming. We will discuss unique case studies that are clinically applicable in daily practice.

Presenter Bio:

Dr. Dansie received a B.S. degree in audiology and speech-language pathology (communicative disorders) from Brigham Young University in 2006. He obtained a Doctorate of Audiology (Au.D.) degree at Utah State University in 2010. He is a certified member of the American Speech-Language Hearing Association, a fellow of the American Academy of Audiology, a member of the American Cochlear Implant Alliance, and a member of USHA. He earned a graduate degree emphasis at USU in pediatrics and auditory learning and spoken language development. Dr. Dansie has specialized training and experience in advanced diagnostic audiometry, pediatrics, electrophysiology, cochlear implants, temporal bone implants, amplification, as well as most areas of audiology. He specializes in practice development. He has worked in a large private practice Neuro-Otology/Otology/ENT office for the last 8 years.
Advanced Diagnostics and Audiometric Testing to Aid In Differential Diagnosis

- Review of the anatomy of the ear.
- Abnormalities of the middle and inner ear structures
  - Enlarged Vestibular Aqueduct
  - Otosclerosis
  - Superior Canal Dehiscence

Differential diagnosis—Use of acoustic reflexes
- Beyond the Audiogram, what more can we do.
- Patulous Eustachian Tube Testing
- Eustachian Tube Function Testing
- Case Studies Throughout Presentation

Anatomy Of The Ear

- External auditory canal
- Middle ear
- Inner ear
- Eustachian tube
- Vestibular nerve
- Cochlea
- Facial nerve
- Semicircular canals
- Auditory nerve
- Tympanic membrane
- Eustachian tube
- Malleus
- Incus
- Stapes

1. Eardrum
2. Malleus
3. Incus
4. Stapes
5. Semicircular canals
6. Auditory nerve
7. Facial Nerve
8. Vestibular nerve
9. Cochlea
10. Eustachian tube
Large Vestibular Aqueduct (LVA)

- Congenital or manifests early in life.
- Usually bilateral
- Asymmetric
- Fluctuating and progressive
- Sudden onset
- Sensorineural, conductive or mixed in nature
- Often times has an air bone gap in the absence of ME pathology
- Often reported to be precipitated by head trauma
- Can have Vestibular symptoms and signs
How is LVA diagnosed?

- LVA is diagnosed by radiological imaging such as CT or MRI.
- Two most common criteria are “Cincinnati criteria” or “Valvasori criteria”
- Cincinnati Criteria = greater than 1.5mm at the midpoint of VA
- Valvasori criteria = equal or greater than 1.2mm at the midpoint and 1.3mm at the operculum.

Studies have shown that the Cincinnati criteria is more sensitive than the Valvasori criteria in diagnosis of EVA.

Normal opercular and midpoint VA measurements. The width of the VA at the operculum (*) is 1.4 mm (long white line), and the width at the midpoint (short white line) is 0.4 mm in this temporal bone. The crus commune (CC) is in the axial plane of this VA.

Imaging for EVA

- When imaging is performed EVA accounts for up to 37% of children with unexplained sensorineural hearing loss (Greinwald et al. 2013)
- Does it change what you might fit? (Headroom/progressiveness)
- Does it change how you might counsel the parents or the patient?
  - Progressive nature
  - Avoid contact sports or head trauma?
  - Can change suddenly—Understanding possible options and outcomes such as a CI
- Associated disorders with EVA
  - Pendred syndrome (SLC26A4 “PDS” gene) (1/3 of patient with EVA)
  - Branchiootorenal Syndrome (kidney problems)
  - Inner ear malformations (Mondini malformation)
  - Waardenburg’s syndrome
Development of EVA

Controversy

- Post Natal Development: Some studies suggest like one by Dr. Mark Pyle in 2000, that unlike the inner ear which is thought to be fully developed at birth, the vestibular aqueduct may continue to mature in size, even post-natally.

- Complete In utero: There is evidence imaging data to suggest that there is no change in vestibular aqueduct development following birth, when there is no difference in size when comparing the adults and children.

- Arrested Development: Some think that the vestibular aqueduct is initially wider and throughout fetal development, it elongates and takes on a J shape. With EVA it is possible that it remains wide, does not elongate, this may occur around 5 weeks gestation.

Prevalence Statistics of EVA

- Prevalence is estimated to be anywhere from 1%-14% of sensorineural hearing loss.

- Bilateral to unilateral ratio is 2:1 (bilateral is more common)

- Female to male ratio is 3:2 (more common in females)

- Can occur in isolation or in conjunction with other congenital disorders such as, Pendred syndrome (SLC26A4 gene) which can have thyroid problems such as hypothyroidism and a goiter, CHARGE syndrome, Branchiootorenal (BOR) syndrome, Waardenburg’s syndrome.
Clinical Presentation of EVA

- Failed a school screening
- Reduced Responsiveness (parent report)
- Speech and Language concerns
- Vestibular symptoms
- Due to school screenings, children that were not identified at birth will come in around the ages of 3-6 years of age.

Audiological Test Battery

- Tympanometry
- Acoustic Reflexes
- Pure tone audiometry
- Speech perception testing: SRT and WRS
- Otoacoustic emissions (OAEs)
- Vestibular Evoked Myogenic Potentials (VEMP)
- Bone conduction testing at 250 Hz and below 0dBHL
Conductive, Mixed, and Sensorineural have all been reported. Conductive and mixed component is likely to occur in the low frequencies, such as 250, 500, and even 1000Hz. Bone conduction can be very good and be at the lowest end of normal (i.e. -10dBHL). This may be missed if you are not testing below 0dBHL. Often times it will present as a sensorineural components in the highs. Hearing loss can occur progress with fluctuations, gradual or suddenly. We will likely see the hearing loss shift from conductive or mixed to sensorineural.

Most common configuration is:
- down sloping
- flat
- reverse cookie bite

Degree of Loss can be all over the audiogram:
- Anywhere from normal to profound
- Inner ear anomalies may affect the progression and degree of loss.
- Can be profound at birth or stable into adulthood (30-40% of patients)
  - 60% of patients can have a gradual progression or sudden changes
Why have a screener on a diagnostic bridge?

Why is there a screening option on a clinical middle ear analyzer?

- Fast method to evaluate middle ear status
- Determine if more testing is appropriate
- Infants, children and difficult to test patients

Further Testing of Reflexes
Case Study #1

- 2 year old male
- Was originally referred by the state school for the Deaf into our clinic for a known unilateral hearing loss.
- Parents had previously chosen not to pursue amplification at birth, but was referred to our clinic due to less consistent responses and possible progression of the hearing loss ear. Possible history of middle ear fluid.
- Some speech and language concerns at the time of referral.

TEST RESULTS

On both tests the right tympanogram was within normal limits, suggesting normal middle ear function, and the left tympanogram indicated negative pressure in the middle ear cavity.

Acoustic reflexes were present within normal limits for the left ear, but acoustic reflex testing was not completed for the right ear due to the probe constantly being pulled from the ear.

The right ear passed the distortion product otoacoustic emissions (DPOAE) screening, but the left ear failed the screening.

Right ear air conduction testing revealed normal hearing sensitivity across all tested pitches. Left ear air conduction testing revealed normal hearing sensitivity at 500 Hz dropping to minimal, mild and moderate-severe hearing loss in the mid and high pitches. These results are possibly "minimal response levels," suggesting that actual thresholds are possibly better than indicated.

The speech audiometric thresholds (SRT’s) were 15 dB HL for the right ear and 20 dB HL for the left ear.

RECOMMENDATIONS:
1) Hearing aids were briefly discussed. If the results are accurate, a hearing aid for the left ear may be beneficial.
   However, results are possibly influenced by PTA’s and middle ear involvement (as seen by the tympanogram testing suggesting that the left ear has a slightly retro-reflecting.
2) PT may benefit from a visit to his physician or an ENT specialist if fluid in the left ear is suspected.
3) Routine hearing testing is recommended every 1 to 3 months, or before, if concerns arise regarding his hearing or speech and language development. An appointment for the hearing test was made for December 2, 2016, at 9 am.

Please feel free to call with questions.
Case Study #1 Ipsilateral Reflexes 500 Hz

Case Study #1 Ipsilateral Reflexes 1000 Hz
Case Study #2 Bilateral EVA

Case Study #2 Adult EVA - 500 Hz Reflexes
Case Study #2 Adult EVA- 1000 Hz Reflexes

Case Study #2 Adult EVA- 2000 Hz Reflexes
Case Study #2 Adult EVA-Audiogram

Adult EVA 500 Hz acoustic reflexes
Adult EVA 1000 Hz acoustic reflexes

Adult EVA acoustic reflexes
Otosclerosis

Otosclerosis Case Study #3
Superior Canal Dehiscence

Symptoms include:
- Autophony
- Dizziness and vertigo
- Tulio's Phenomenon
- Pulse synchronized nystagmus
- Hyperacusis
- Conductive hearing loss
- Aural fullness
- Pulsatile tinnitus
- Foggy sensation
- Fatigue
- Headaches/Migraines
- Tinnitus

Case Study #4

- 32yo Female

- Reports pulsatile tinnitus, worse with exercise and/or increased heart rate.
- Hears heartbeat in left ear during strenuous workouts.
- Also reports nystagmus following exercise, when heart rate is slowing
Case Study #4

CT SCAN LEFT SCC
7 year old male

Was referred into our practice by his pediatrician for concerns about possible bilateral cholesteatomas.

Parent report indicates that he passed on his newborn hearing screening.

Pt was born at 34 weeks, but went home with his mother at discharge from hospital.

Speech and language development are normal.

- Manubrium of the Malleus
- Thickening of the TM
- Retraction Pocket
- Head of the stapes
Case Study #5 – 7 Year old male

- Patient received PE tubes bilaterally
- Right ear hearing was very similar, but tested with inserts
- Left ear hearing decreased dramatically from a normal to mild SN loss to a near maximal conductive loss.
- WRS was reported to be 80% at 90dBHL, but reliability was deemed poor for the left ear.
- An ABR was recommended by the audiologist
Later…..
Tubes have fallen out, still some negative pressure, but better than preoperative
Right has a slight to normal sensorineural hearing loss, with good WRS
Left ear has a moderately severe to slight mixed loss, but when masked WRS is very poor (8%).
CT showed only 2 turns of the cochlea, bilaterally
Vestibular apparatus was unremarkable bilaterally
Vestibular aqueduct was unremarkable bilaterally

Immittance Testing for Cochlear Implants
Immittance testing for CI programming

- When patients have trouble doing loudness scaling to set loud levels (M/C) levels
- When the patient has a hard time judging between loudness and pitch
- When the patient is not capable of giving loudness ratings (young children or multiply involved patients)
- Tympanograms and Electrical Stapedial Reflex Testing

ESRT Set up

- Requires patient to sit still for 5-10 min
- Requires patient to allow immittance probe to stay in the ear.
- Does not require any active participation of the patient to use for programming
- Patient can watch a movie, or play a game on a phone or iPad.
Electrical Stapedial Reflex Testing – CI Software

Electrical Stapedial Reflex Testing – GSI TympStar Pro
6 year old patient came in after referring on a school screening
- Reportedly passed newborn hearing screening
- Audiometric testing at age 6 showed hearing within normal limits in the right ear and no response audiogram in the left ear.
- Pt had developed normal speech and language, the parents had not been aware of any hearing loss
- Failed trials of CROS and Softband Baha
- Parents didn’t want to do a Baha Attract or Connect
- When the Baha SoundArc was released, he was invited back in to do some testing.
- QuickSIN testing showed measurable improvement in background noise
- Pt was fitted with his own SoundArc Baha device.
Patulous Eustachian Tube

- Patient reports distorted autophony, echoing when talking, wave-like sounds, sensation of aural fullness.

Can occur after:
- weight loss
- Pregnancy
- Neurological disorders such as: muscle atrophy, stroke, multiple sclerosis, motor neuron diseases
- Diuretics

Patulous Eustachian Tube set up......

- Set the immittance bridge to Decay (external or contralateral stimulus)
- Put the immittance probe in the ear
- Have the patient plug the contralateral nostril
- Have the patient breath deeply through the nose
Patulous Eustachian Tube Patient Set Up

Patulous Eustachian Tube Testing – GSI TympStar Pro
Patulous Eustachian Tube Testing Video

Eustachian Tube Function (ETF) Testing
Eustachian Tube Function (ETF) Testing

- Have a cup of water available
- Pressurize the tympanometer (ETF)
- Tympanometer will give instructions when to drink, (after each tymp)
- (pressure moves to +400 daPa, -400 daPa)
- Total shift in peak pressure should be 15-20 daPa

Hearing Aid Case Study #1

- Bimodal patient came in for a cochlear implant consult for his second ear
- Reported that when he uses his tractor, it gets extremely/uncomfortably loud, as if noise reduction was not working or kicking in.
Use of Real Ear and Test box measures for troubleshooting

HA case study #2

- 56 year old male had been fitted with a custom made for iPhone product.
- Overall a very pleasant patient but had one major complaint with his custom hearing aids.
- He had demoed RICs and enjoyed the connectivity and sound quality but with the custom product he reported that he would hear the hearing drop significantly in the high frequencies after about 20-30 seconds after starting up or switching programs, but not when hooked to the computer.
- The audiologist had done many things to try to improve this, turning off noise reduction, directionality, any advanced features, performed speech mapping, had the hearing aids remade, created new Noah files x3 per the manufacturer recommendations. None of this helped.
- The audiologist was at a loss, he was going to send the hearing aids back for a second remake or try to talk the patient into a style change, since he did not have this same problem with the RICs.
- The patient had been in a total of about 14 visits in a 5-6 month period.
HA case Study #2

- We decided to run real ear again but first not when it was hooked to the computer (since the problem didn’t occur when hooked up).
- Sure enough we would see a decrease in the high frequencies after about 30 seconds.
- Solution?

Conclusion

- Use our equipment to help guide us and make correct decisions
- Quick, easy, and billable tests can help identify disorders or at least raise red flags to catch disorders and aid in good clinical decision making
- When we get something that doesn’t make sense, trust your instincts
- Make sure we use good, sound testing principles to not miss something bigger.
- Don’t be afraid to get creative with the tools that we have in our clinic to help identify and resolve patient complaints in the clinic.
Questions?

References ....

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