Clinical Updates in Vestibular
Meniere’s, Vestibular Migraine, CSD, and High Frequency/Speed Head Movement Testing

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Clinical Updates in Vestibular
Meniere’s, Migraine, Psychological Factors
Index Case

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**43 y.o. Female**

- **CC – constant non-vertiginous dizziness exacerbated with head / visual motion, visual complexity, reading plus spontaneous vertigo events**

- **Past Medical Surgical History**
  - Prior and active history of migraine
  - 1999 fluctuant & progressive Hearing loss on the left
  - 2000 began spontaneous vertigo events – 1-2 hours multiple per mo

- **Past Medical Surgical History – cont**
  - Dietary & diuretic did not control
  - 2004 left endolymphatic sac decompression – good control
  - 2007 spells returned increasing frequency with falls in 2008 – between spells non-vertiginous sx provoked by visual patterns, visual motion and visual complexity. The spontaneous vertigo began with a 4 day spell.
43 y.o. Female

- Past Medical Surgical History – cont
  - Left Vestibular Nerve section 2008
  - Good control of spontaneous events with reduction in visually provoked other symptoms
  - 1 year later spontaneous events began to return – suspicion of incomplete nerve section
  - In 2009 50% of spontaneous events were with a following focal headache – the other 50% were with light and odor sensitivity. Offered Labyrinthectomy but cancelled.

43 y.o. Female

- Current symptoms at time of 2010 evaluation
  - Sensation of self motion (vertigo in her head) that is 24 / 7
  - Spontaneous exacerbations
  - Exacerbations with head movement, visual motion, visual complexity and repeated noise
  - Falls but not of Tumarkin Crisis description
  - Continues with left hearing fluctuations
43 y.o. Female

- Laboratory & direct examination
  - MRI normal for CPAs and IACs
  - Highly positive for anxiety and depression via the HADS
  - No abnormal nystagmus on any condition tested
  - Head thrusts, warm and ice water irrigations and rotary chair indicated severe left peripheral hypofunction in a partial state of compensation
  - Mild central findings with normal MRI likely migraine related
43 y.o. Female

• Seen by members of the dizziness & balance disorders team
  • Dr. Neff – Neurotologist
  • Dr. Eggers – Neurologist
  • Dr. Staab – Psychiatrist

Meniere’s Disease: Focus on Treatment

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Presentation

• Fluctuating low tone sensorineural hearing loss
  • Can be flat, sudden, progressive, or profound loss.

• Repetitive vertigo- rotary, 20 min.- < 24 hours
  • Can have dysequilibrium
  • Other descriptions of environmental movement
  • Must be spontaneous onset- not motion provoked

Presentation

• Aural fullness- pressure
• Tinnitus- usually low tone, roaring; can have other descriptions
• Timing- the ear symptoms usually precede the vertigo by a short amount of time- not always
• Usually unilateral; can be bilateral
## Diagnostic Criteria

### Certain Meniere’s Disease
1) Definite Meniere’s Disease plus histopathologic confirmation

### Definite Meniere’s Disease
1) Two or more definitive episodes of spontaneous vertigo lasting > 20 minutes
2) Audiometrically documented hearing loss on at least one occasion
3) Tinnitus or aural fullness in the suspected ear
4) Other causes excluded

### Probable Meniere’s Disease
1) One definitive episode of spontaneous vertigo lasting > 20 minutes
2) Audiometrically documented hearing loss on at least one occasion
3) Tinnitus or aural fullness in the suspected ear
4) Other causes excluded
**Diagnostic Criteria**

**Possible Meniere’s Disease**

1) Episodic vertigo of the Meniere’s type without documented hearing loss

   OR

2) Sensorineural hearing loss, fluctuating or fixed, with disequilibrium but without definitive episodes

3) Other causes excluded

---

**Treatment Overview**

• No therapy has been found to help:
  • Hearing loss
    - IT steroids?, endolymphatic sac surgery?
    - Hearing aids and cochlear implants
  • Tinnitus
  • Aural fullness
Treatment Overview

- Lot of variability in approach
  - No randomized, blinded, placebo controlled trials or clear cut “best practices” studies
  - Physicians often feel that effectiveness in “their hands” is under or overstated by the medical literature
- Reliant on common sense approach

Non-ablative Treatments

- Medical and diet therapy
- Transtympanic steroid treatments
- Meniett device™ (Medtronic, Xomed)
- Endolymphatic sac surgery
Ablative Therapies

- Transtympanic gentamicin injections
- Vestibular nerve section
- Labyrinthectomy

Medical Therapy

- Low salt diet <1500 mg/day Na⁺
  - Amount controversial
- Diuretics
  - Dyazide (triamterene/HCTZ)- one pill each AM- not with sulfa allergy
- Regular sleep, regular exercise, stress reduction, avoid caffeine and nicotine
Medical Therapy

• Oral steroids
  • Yes- if thought to be autoimmune
  • Role in standard Meniere’s not clear

• Antihistamines, herbal therapies, hyperbaric oxygen, vitamins, acupuncture, chiropractor etc..
  …no proof that they work!

Medical Therapy

• Concomitant migraine
  • Migraine diet
  • Important to try daily migraine prophylaxis first- no irreversible risk to hearing

• Concomitant chronic subjective dizziness (CSD)
  • Important to treat along with Meniere’s disease or outcomes likely to be poorer
Transtympanic IT Steroids

- Not standard agreement on which steroid to use, method of delivery, or dosing schedule
- Delivered to the middle ear via injection through the eardrum.
- Absorbed into inner ear via round window membrane

- 3 doses of 24mg/cc dexamethasone given 2-3 weeks apart.
- 5-10% chance of perforated eardrum with conductive hearing loss
- 1% chance of infection - otitis media or draining ear
Meniett™ Device

- Device sends low-pressure pulses (35 cm H₂O) transmitted through a tube in the eardrum to the round window membrane
  - Use for 6-12 weeks; 2-3 times/day
- Helps inner ear absorb excess fluid and function more normally
  - This is all an educated hypothesis and not fact

- Many insurances will not cover $3000-3500 cost of device.
- Tube has to be placed into ear
- Not utilized by many ENTs—controversial effectiveness

Hain TC. http://www.dizziness-and-hearing.com/images/Meniett.jpg
Endolymphatic Sac Surgery

- Surgery done to remove mastoid bone over the endolymphatic sac and to place a silastic shunt into sac
- Unclear mechanism—shunt excess fluid? Damage the sac?
- Controversial effectiveness (placebo?); many ENTs still perform

Endolymphatic Sac Surgery

- Controversial whether “shunt” necessary—just decompress sac?
- 1% risk of facial paralysis, 5-10% risk of prolonged dysequilibrium, 1-2% risk of CSF leak
IT Gentamicin

• Administered similar to steroid injection

• Mechanism
  • Selectively vestibulotoxic to hair cells in injected ear- “controlled vestibular ablation”
  • Compensation occurs if contralateral ear has intact vestibular system

• No general consensus on the best dosing schedule, method etc.

IT Gentamicin

• Can not do complete vestibular ablation bilaterally- usually unilateral disease with exceptions
  • Titration method (40mg/cc)- inject every 6 weeks; max of 3 injections
  • Base treatment end-point on vertigo control not testing

• 5% chance of worsened balance with walking, 1% chance of perforated eardrum
Vestibular Nerve Section

- Craniotomy with section of the vestibular nerve with attempted preservation of the cochlear nerve
- Can not be done bilaterally - disabling oscillopsia
- Not uncommon to have difficulty distinguishing vestibular from cochlear nerve fibers - unintended postoperative hearing loss

Vestibular Nerve Section

- It is invasive
  - 5% risk of CSF leak, facial paresis, or worsened balance with walking
  - 1% risk of meningitis, intracranial hemorrhage, permanent facial paralysis, or stroke
  - 5-10% prolonged or permanent post-craniotomy headaches
    - Problem - population with migraines
Labyrinthectomy

- Surgery done from behind ear
  - Remove mastoid bone, all 3 semicircular canals, utricle and saccule.
  - Hearing is intentionally sacrificed in the operated ear in all cases
  - Best vertigo control rate

- Still has risks - 5% worsened balance with walking, 1% risk of facial paralysis or CSF leak
- Can not do bilaterally due to disabling oscillopsia and profound bilateral hearing loss
Vertigo Control

- Medical therapy 50%
- Meniett™ device 50%
- IT steroids 50%
- Endolymphatic sac surgery 65-70%
- Gentamicin injection 80-90%
- Vestibular nerve section 85-90%
- Labyrinthectomy 95%

Risk for Treatment Induced Hearing Loss

- Medical therapy ~ 0%
- IT steroids <5%
- Meniett™ device <5%
- ELS surgery 10-15%
- IT gentamicin 15-20%
- Vestibular nerve section 15-20%
- Labyrinthectomy 100%
Conclusion

- Approach depends on patient goals, patient risk assessment, hearing level, and common sense
- Hopefully a better understanding of Meniere’s disease pathophysiology will lead to improvements in future therapies

Vestibular Migraine Update

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Ménière’s Disease & Migraine

• Association postulated by Ménière 1861

• About half of Ménière’s Disease patients…
  • Meet IHS criteria for migraine
  • Always have at least 1 migraine symptom during Ménière’s attacks
    Migrainous headache 28%, photophobia 46%, aura 10%

• Mild fluctuating cochlear symptoms are common in vestibular migraine (binaural tinnitus, fullness)

Radke et al. Neurology 2002
Johnson. Laryngoscope 1998
Olsson. Laryngoscope 1991

Ménière’s Disease & Migraine

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Johnson. Laryngoscope 1998
Olsson. Laryngoscope 1991
**Vestibular Migraine**

The interrelations of migraine, vertigo, and migrainous vertigo

- Vertigo is 3 times more common in migraineurs than in tension headache patients
- Migraine is overly common among vertigo patients
- 61 to 87% of patients with chronic recurrent attacks of vertigo without otologic symptoms or identifiable cause have migraine

References:
- Kayan & Hood. Brain 1984
- Neuhauser et al. Neurology 2001
- Savundra et al. Cephalalgia 1997
- Cho et al. Cephalalgia 2009

**Vertigo With Migraine**

- Vertigo is 3 times more common in migraineurs than in tension headache patients
- Migraine is overly common among vertigo patients
- 61 to 87% of patients with chronic recurrent attacks of vertigo without otologic symptoms or identifiable cause have migraine

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- Kayan & Hood. Brain 1984
- Neuhauser et al. Neurology 2001
- Savundra et al. Cephalalgia 1997
- Cho et al. Cephalalgia 2009
Pathophysiology
Hypotheses

- Central and peripheral vestibular involvement
- Spreading depression affecting brainstem or cortical vestibular structures
- Vasospasm of internal auditory artery
  - Accounts for peripheral vestibulopathy and hearing loss?
- Activation of locus ceruleus and dorsal raphe projections to vestibular nuclei
- CGRP release in vestibular nuclei and labyrinth
- Defects in ion channels (overlap with EA-2, FHM)
  - CACNA1A, ATP1A2, SCN1A, CACNB4 negative
  - Genome-wide linkage screen: 22q12 linkage in BRV

Diagnostic Criteria for VM

Proposed by Neuhauser et al. (2001 and 2004)

Definite vestibular migraine

A. Recurrent episodic vestibular symptoms of at least moderate severity
B. Current or previous history of migraine according to the criteria of the International Headache Society
C. One of the following migrainous symptoms during at least two vertiginous attacks: migrainous headache, photophobia, phonophobia, visual or other auras
D. Other causes ruled out by appropriate investigations

>5 headaches, 4-72hrs
- N/V or photo/phonophobia
- 2 of: unilateral, pulsatile, moderate-severe, aggravated by motion

Furman et al. JNNP 2005
Cutrer & Baloh. Headache 1992
Kirchmann et al. Neurol 2006
Lee et al. Hum Mol Gen 2006
Ahn et al. Neurosci Lett 2009
Vestibular Migraine

- Most common cause of recurrent spontaneous episodes of vertigo
  - 1% prevalence among general population in study of almost 5000 adults
- “Migrainous vertigo” or “migraine-related dizziness”
- Variable manifestations (may mimic Ménière’s)
- Diagnosed by the company it keeps (no test)
  - Young/middle age, female predominance
  - History of motion sickness
  - Family history of migraine or vertigo
  - Migraine triggers

VM Symptoms

- Vertigo episodes may last minutes, hours, or days
  - (Few other conditions cause vertigo episodes lasting days)
- Variable temporal relationship between vertigo and headache
  - 50-70% report that some of their vertigo attacks are during or followed by headache
- Minor auditory Sx: phonophobia > tinnitus > hearing loss
- Photophobia, osmophobia, visual aura

References:
- Neuhauser et al. Neurology 2006
- Dieterich & Brandt. J Neurol 1999
- Cutrer & Baloh. Headache 1992
- Brantberg et al. Acta Otol 2005
- Harker & Rassekh. Laryngoscope 1988

- Dieterich and Brandt. J Neurol 1999
- Kayan and Hood. Brain 1984
- Cutrer. Headache 1992
VM Examination

- Generally normal between attacks
- Exam during attack
  - Abnormal nystagmus in 70% (central>peripheral)
  - Spontaneous, gaze-evoked, sustained positional
  - Most have impaired gait/balance but can still walk

Vestibular Testing in VM

- Minor abnormalities are common (as in migraine w/o vertigo), but no specific testing abnormality for VM
- Central and peripheral
  - Unilateral (mild) caloric weakness in 10-20%
  - Rotary chair directional preponderance in 20%
  - Elevated visually-enhanced VOR?
  - Mild spontaneous or sustained positional nystagmus
  - Central ocular motor deficits
    - Smooth pursuit, VOR suppression
  - Posturography usually normal
  - Audiometry: non-fluctuating/progressive SNHL <10%
  - VEMPs: reduced amplitudes, increased latency?

- Testing may trigger a migraine, motion sickness
Red Flags That it’s Not Migraine

- New onset spontaneous vertigo in elderly
  - Vertebrobasilar disease
- Significant aural Sx with vertigo (monaural hearing loss, roaring tinnitus)
  - Ménière’s Disease
- Single prolonged episode of spontaneous vertigo with residual imbalance
  - Vestibular neuritis
- Exclusively brief episodes of positional vertigo
  - BPPV
- Progressive unilateral HL & ataxia
  - Vestibular schwannoma

Migraine Management

- Avoid triggers, sleep hygiene, exercise, etc
- Acute attacks > 30 min
  - Vestibular suppressants
    - Meclizine, promethazine, diazepam
  - Triptans?
- Prophylaxis if frequent or severe attacks
  - No controlled studies; choose based on side effects
    - Nortriptyline, topiramate, gabapentin, propranolol, verapamil, venlafaxine
- Address co-morbid conditions
Comorbid Chronic Subjective Dizziness

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Chronic Subjective Dizziness

• Persistent dizziness or unsteadiness
  • Lightheadedness, heavy headedness, “swimming”

• Hypersensitivity to motion stimuli
  • One’s own movements
  • Motion cues in the environment

• Provocation with visual challenges
  • Active visual environments
  • Visually demanding tasks

Staab and Ruckenstein, Arch Oto-HNS, 2005, 2007
Comorbidity is Common

507 diagnoses in 410 Patients

- Meniere's
- BPPV
- PVD/VN
- CVD
- CSD
- Migraine

Vestibular Symptom Phenotypes

1. Episodic vertigo
   Spells of rotation or lateral motion
   Self or surround

2. Episodic unsteadiness
   Spells of swaying or rocking
   Self or surround

3. Chronic unsteadiness
   Persistent swaying or rocking
   Impairment in sense of space
   Heavy-headed, lightheaded, full/foggy
Episodic Vertigo & Migraine

• Rates of migraine
  Tertiary dizziness clinic (N=200)
  Patients with episodic vertigo

<table>
<thead>
<tr>
<th></th>
<th>Vertigo</th>
<th>Public</th>
<th>RR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>38.0%</td>
<td>15.0%</td>
<td>2.5</td>
</tr>
</tbody>
</table>

Neuhauser, et al., Neurol, 2001

Chronic Dizziness & Anxiety

• Rates of migraine and anxiety disorders
  • Tertiary dizziness clinic (N=345)
  Patients with chronic, non-vertiginous dizziness

<table>
<thead>
<tr>
<th></th>
<th>Dizzy</th>
<th>Public</th>
<th>RR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>16.5%</td>
<td>15.0%</td>
<td>1.1</td>
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<tr>
<td>Anxiety disorders</td>
<td>79.1%</td>
<td>18.1%</td>
<td>4.4</td>
</tr>
</tbody>
</table>

Staab & Ruckenstein, Arch Oto HNS, 2007
Our Patient – Symptoms & Diagnoses

• Episodic vertigo, unsteadiness
  ✓ Ménière's disease
  ✓ Migraine
  ✗ CSD

• Chronic (daily) dizziness
  ✗ Ménière's disease
  ? Migraine
  ✓ CSD

Anxiety Complicates Med/Surg Tx
Transtympanic gentamicin for Ménière’s disease

Boleas-Aguirre et al., Laryngoscope, 2007
### Behavioral Treatment Options

- **Medication**
  - SSRI – fluoxetine (Prozac) group + migraine prophylactic medication
  - SNRI – venlafaxine (Effexor) group
  - Tricyclic antidepressant – nortriptyline group
- **Rehabilitation**
  - VBRT – habituation style
- **Psychotherapy**
  - Cognitive behavior therapy (CBT)

### SSRI Treatment of Chronic Dizziness

<table>
<thead>
<tr>
<th>Authors</th>
<th>Study Type</th>
<th>Tx Wks</th>
<th>Medication</th>
<th>N Total</th>
<th>Diagnostic Group</th>
<th>N per Group</th>
<th>Responders n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Staab, et al., 2002a</td>
<td>retrospective case series</td>
<td>20 various SSRIs</td>
<td>60</td>
<td>CSD+anxiety CSD</td>
<td>31 14</td>
<td>27 (87%) 11 (79%)</td>
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<tr>
<td>Horii, et al., 2004</td>
<td>prospective open label</td>
<td>8 paroxetine 20 mg/d</td>
<td>47</td>
<td>PVD+dep PVD CSD+dep CSD</td>
<td>12 17 6 12</td>
<td>12 (100%) 0 (0%) 5 (83%) 0 (0%)</td>
<td></td>
</tr>
<tr>
<td>Staab, et al., 2004</td>
<td>prospective open label</td>
<td>16 sertraline 50-200 mg/d</td>
<td>20</td>
<td>CSD+anxiety CSD</td>
<td>11 4</td>
<td>8 (73%) 3 (75%)</td>
<td></td>
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<tr>
<td>Simon, et al., 2005</td>
<td>prospective open label</td>
<td>12 fluoxetine 20-60 mg/d</td>
<td>5</td>
<td>CSD PVD</td>
<td>2 3</td>
<td>2 (100%) 0 (0%)</td>
<td></td>
</tr>
<tr>
<td>Horii, et al., in press</td>
<td>prospective open label</td>
<td>8 fluvoxamine 200 mg/d</td>
<td>60</td>
<td>PVD+anx/dep CSD+anx/dep</td>
<td>19 22</td>
<td>sig↓ anx/dep &amp; dizziness in both groups</td>
<td></td>
</tr>
</tbody>
</table>
SNRI – Migraine, Anxiety, Dizziness

- 32 patients -- 8 weeks of treatment with venlafaxine XR (mean dose = 225 mg/d)
  - Dizziness - much or very much improved
  - Headaches - reduced by 50% in frequency/severity

<table>
<thead>
<tr>
<th>Dizzy patients</th>
<th>N</th>
<th>Dropouts</th>
<th>Completers</th>
<th>Responders</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine with anxiety</td>
<td>20</td>
<td>6</td>
<td>14</td>
<td>13</td>
</tr>
<tr>
<td>Migraine alone</td>
<td>12</td>
<td>2</td>
<td>10</td>
<td>4</td>
</tr>
</tbody>
</table>

Intent to treat: 65% vs 33%, X²=3.0, p < 0.10  
Observed cases: 93% vs 40%, X²=7.9, p < 0.01

Staab, JVR, in press

VBRT and CBT for Chronic Dizziness

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Authors</th>
<th>Study Type</th>
<th>N</th>
<th>Study Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>VBRT</td>
<td>Jacob, et al., 2001</td>
<td>Pilot Uncontrolled</td>
<td>9</td>
<td>2 weeks of self-exposure exercises, then 8-12 weeks of therapist-directed VBRT.</td>
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<tr>
<td></td>
<td>Yardley, et al., 2001</td>
<td>Parallel group Randomized</td>
<td>76</td>
<td>33 patients treated with self-exposure exercises versus 43 untreated controls.</td>
</tr>
<tr>
<td></td>
<td>Pavlou, et al. 2004</td>
<td>Parallel group Randomized</td>
<td>40</td>
<td>20 patients treated with VBRT versus 20 treated with VBRT plus desensitization with a visual motion simulator.</td>
</tr>
<tr>
<td>CBT</td>
<td>Holmberg, et al., 2006</td>
<td>Parallel group Randomized</td>
<td>31</td>
<td>16 patients treated with CBT versus 15 treated with self-exposure exercises.</td>
</tr>
<tr>
<td></td>
<td>Johansson et al., 2001</td>
<td>Pilot Wait-list Control</td>
<td>19</td>
<td>9 elderly patients treated with CBT plus VBRT versus 10 patients on a waiting list.</td>
</tr>
</tbody>
</table>
Behavioral Treatment Plan

- Medication – *sequential trails*
  - SSRI – fluoxetine (Prozac) group
  - + migraine prophylactic medication
  - SNRI – venlafaxine (Effexor) group
  - Tricyclic antidepressant – nortriptyline group
- Rehabilitation – *definitely*
  - VBRT – habituation style
- Psychotherapy – *possibly*
  - Cognitive behavior therapy (CBT)

Clinical Updates in Vestibular

Meniere’s, Migraine, Psychological Factors

Summary

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43 y.o. Female

- Seen by members of the dizziness & balance disorders team
  - Dr. Neff – Neurotologist
    Left Meniere’s not the likely cause of sx; Migraine related dizziness; Chronic Subjective Dizziness (CSD)
  - Dr. Eggers – Neurologist
    Left Meniere’s not likely cause of sx; migrainous vertigo; CSD
  - Dr. Staab – Psychiatrist
    CSD; migrainous vertigo; Meniere’s not active; Major depression; Posttraumatic stress disorder; psychological factors affecting medical condition

43 y.o. Female

- Recommendations
  - Maintain current medication for depression
  - Prophylactic migraine therapy 1st with non-antidepressant (e.g. topiramate or gabapentin)
  - Habituation exercises for desensitization for visual motion & complexity +/- Cognitive behavioral therapy
  - No further aggressive treatment for Meniere’s
Combined Diagnoses

- Meniere’s < 0.25% of general population
- Migraine headaches 14-17% of general population
- Generalized Anxiety < 2%
- Migraine in Meniere’s as high as 50%
- Anxiety with Migraine up to 30%

Treatment Challenge

- What aspect seems to be the greatest impact on the functionality of the patient?
  - In this case Migraine and CSD
  - In others Meniere’s (other peripheral disorders) may carry the greatest impact
Probe Microphone Measurements: Best Practice or Optional?

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“Although it is true that mere detection of a sound does not ensure its recognition, it is even more true that without detection the probabilities of correct identification are greatly diminished.”

(Pascoe, 1980)
Without Audibility

- Better have good eyes in order to utilize visual cues
- Better have good cognitive function to use context and other cues
- Better prepare to use a lot of cognitive effort
- Using eyes and brain plus having audibility gives the hearing-impaired person the best chance at speech intelligibility and better quality of life

What Makes a Successful Hearing Aid Fitting?

- Many things, some unrelated to audibility, such as:
  - Patient motivation is good
  - Appropriate expectations
  - Style of aid is acceptable to the patient, appropriate to the hearing loss, and is comfortable
  - Patient can insert the aid and change the battery
  - Good counseling and follow up
  - Patient is committed to adjustment
Assumption: You’re Good, You’ve Done All That

• Focus on what the instrument needs to do acoustically
• And that is also determined by YOU!
• You have the opportunity to do this adjustment
  • With precision
  • For each individual patient
  • To conform to “best practices”

What Do You Want The Hearing Aid To Do?

• Package the desired signal (speech) into the patient’s residual dynamic range (which will be reduced) so that
  • Speech sounds are audible
  • Average speech levels are comfortably loud

• Allow loud sounds to be loud, but not uncomfortable

• Bottom line: audibility, comfortable loudness, no discomfort for loud sounds
Packaging Speech into the Residual Dynamic Range

Skinner (1979)
Interesting Study in This Regard

25 amplified LTSS 5 dB above threshold and 5 dB below discomfort

Summary

- Package long-term speech spectrum at an appropriate place within the user’s residual auditory dynamic range
- Adjust so that:
  - Soft sounds are soft
  - Normal sounds are comfortable
  - Loud sounds are loud but not uncomfortable
- “Common Sense”
Buy This Argument for Now…
Back to the Clinical World

- How are you actually setting the HA gain?
  - Frequency response?
  - Maximum output?
- Do you know if speech is audible across frequency?
- Do you know where within the DR speech has been placed?
- Are loud sounds not uncomfortable?

If You Don’t Know This…You’re Not Alone…But Shouldn’t You Know It?

- You may have taken…
  - “The Path of Minimal Adjustment”
    - Use the HA manufacturer’s “proprietary algorithm”
    - “First Fit” – assuming that gets you in the ball park
- But does it? We’ll return to that issue in a few minutes
First...How To Think About Audibility: Some Background

• May need a mind shift
  • Think dB SPL in the ear canal
    • Far easier and better than dB HL once you’re used to it
  • Don’t think of hearing aids as improving thresholds… THEY DON’T!
• Thresholds stay elevated
• Hearing aids amplify sounds to a point that they are audible given the impaired threshold

The Basic Framework

SPL in the ear canal, not HL, not free-field SPL

Normal threshold in dB SPL in the ear canal
Average Speech Spectrum at 65 dB SPL for Normal Hearing

Unaided average long-term speech spectrum in ear canal

Speech peaks

Speech valleys

Normal threshold in dB SPL in ear canal

Now Add A Hearing Loss

Thresholds converted to dB SPL in the ear canal
What’s Audible With This Hearing Loss?

Only unaided audible speech energy

What’s Audible for Soft Speech at 50 dB SPL?

Nothing much audible...mainly sees lips moving!
“If People Would Just Speak Up
I Do Fine!”

Some audible info through 2000 Hz

Where Exactly Do We Want the
Hearing Aid To Place the LTSS?

• Certainly package it within the
  reduced residual dynamic range of
  the individual

• But where exactly?
  • DSL (Canada), Version 5.0a
    Want amplified speech bands to
    be comfortably loud
  • NAL-NL1 (Australia, soon NL2)
    Want amplified speech bands to
    be equally loud

• Both have good research support
Where Would DSL 5.0 Put the Average Speech Spectrum?

Where Would NAL-NL1 Put the Average Speech Spectrum?
DSL and NAL Also Provide Maximum Output Targets

DSL MPO targets, RESR

NAL MPO targets, RESR
Placing Speech Within the DR Is Called “Speechmapping”*

- Use probe mic measurements
- Signal is actual speech or speech-like
- Adjust HA with probe tube in the ear until match DSL or NAL targets for
  - Soft speech
  - Average speech
  - Loud speech
  - Maximum output
- Time required two ears: 10-15 min

*Trademarked by AudioScan, 1992

What Do You See With Speechmapping?
Is Speechmapping Necessary? Would “First Fit” Do Same Thing?

- If no verification and adjustment is performed, then you are assuming:
  - Manufacturer’s algorithm has a defined research base
  - “First Fit”
    - Makes speech audible and comfortable
    - Loudness discomfort is avoided
  - Your patient is “average”
  - The NOAH screen is reality

The NOAH Screen for a Moment

Very interesting….
- You “read” the hearing aid and may believe that what’s on the NOAH screen is how that hearing aid is functioning
- Not necessarily….it’s how the average hearing aid circuit of that model should be functioning at the programmed settings
- NOAH is making NO MEASUREMENTS of that specific hearing aid’s performance
• Simple, convincing evidence
  • Connect an ITE and “read” it
  • Now plug the receiver tube with Fun Tac, the aid is functionally dead
  • Read it again, NOAH will give the same result!
• Connect a BTE and “read” it and look at the NOAH 2cc gain
  • Measure the 2cc gain yourself
  • They can be different
  • Intramodel differences

**Actual 2cc Gain Minus NOAH Simulated 2cc Gain**

![Graph showing the comparison between Actual 2cc Gain Minus NOAH Simulated 2cc Gain (dB) and Frequency (kHz)](image)
• If you want to see even more variability:
  • Look at manufacturer’s predicted insertion gain in NOAH software for a hearing aid
  • Now measure insertion gain yourself on the patient
  • Can be very different. Why?
    • Manufacturer’s CORFIG
    • Individual variability
    • Average values are assumed
    • Intramodel differences

---

**Actual REIG Minus NOAH Simulated REIG**

![Graph showing Actual REIG Minus NOAH Simulated REIG](image)

- X-axis: Frequency (kHz)
- Y-axis: Actual IG Minus Simulated IG (dB)
Similar Results from Aarts and Caffee (2005)

The Reality Is….

• NOAH screen is a simulation
• You see average values for that model assuming it is functioning properly
• It may not actually be performing that way
• Predictions of actual performance on your patient may bear no resemblance to how it really works
• Doesn’t show audibility in that person’s residual dynamic range
Back to “First Fit”… What Gain Does It Prescribe?

- Varies dramatically by manufacturer
- Bentler and Chiou
  - Here are the 2cc curves
  - All over the place

What Audibility Might You Get With A “First Fit” Algorithm for a New User?

- Often do it first thing in HA fittings
  - Great education for an Au.D. extern
  - Students often come to 4th year having only done “first fit” and have not done REM to see what happened

- I find the “First Fit” results
  - Bothersome
  -Depressing

- Externs find them “shocking”
Manufacturer “First Fit” for a New User on a Patient of Mine

Aided SII = 22
Predicted Aided CST = 24%

Here’s What I Did

Aided SII = 51
Predicted Aided CST = 94%
Another Manufacturer “First Fit” for a New Patient of Mine

Aided SII = 23
Predicted Aided CST = 33%

Here’s What I Did

Aided SII = 56%
Predicted Aided CST = 96%
What About A Long-Time Hearing Aid User?

• Does amplified speech come close to something like NAL-NL1 targets on a “First Fit” for an experienced user?

• Are the various companies different?

Manufacturer A, Long-Time User, Average Speech Input
Manufacturer B, Long-Time User, Average Speech Input

It’s Not Just Gain…Must Look At The Entire Dynamic Range

- Experienced HA user I had recently
- Just purchased hearing aid elsewhere
- Came to me
  - Everything seems “distorted”
  - Muffled when speaker talks louder and for her own voice
- Listened to aid – she’s right!
Did A Simulated Speechmap

Where The RESR Was Set, Giving Her DR of Only 15-20 dB

Here’s What I Did

I Moved RESR Up To Match DSL Targets
DR Now 30-35 dB, No Distortion and I Verified There Was No Aided Loudness Discomfort
What’s A Better Approach Than First Fit?
What Is “Best Practice?”

- Clear agreement on this question in US and internationally
- AAA Guidelines for Adult Hearing Aid Fitting (2006)
  - “Prescribed gain from a validated prescriptive method should be verified using a probe microphone approach that is referenced to ear canal SPL.”
  - “Output characteristics should be verified using a probe microphone approach that is referenced to ear canal SPL. Determination of audibility at several input levels is the ideal method of verification.”
  • “If probe-microphone measures of real-ear hearing aid performance are not possible, hearing aid performance can be predicted accurately in the real ear by applying age appropriate average RECD values to the measured 2-cc coupler electroacoustic results (Seewald et al., 1999).”

• ASHA (1998) Guidelines
  • “In order to determine how the hearing aids are performing for a given client, probe microphone measures should be made unless contraindicated by physical limitations (e.g., size of ear canal, drainage, excessive cerumen, etc.) These guidelines strongly support the use of real-ear measures, when applicable, as the primary method of verifying the performance of hearing aids.”
International Perspective

Australia

• National Pediatric Protocol for Australia (2010)
  • “Real ear aided gain targets are recognized as providing the most appropriate hearing-aid prescriptions for young children.”

Canada

• Pediatric Amplification Protocol for Ontario Canada Infants (2010)
  • Use Real Ear Coupler Difference or RECD
    • “Approximation of the output of the hearing instrument to the calculated targets is important to ensure that speech is audible and loud sounds are not uncomfortable, across a broad frequency range.”
Recommendation Is Clear: Probe Mic Is Standard of Care

• Are Audiologists doing it?
  • 1995 Hearing Journal survey
    54% of audiologists use routinely
  • 2003 Hearing Journal survey
    37% use routinely
  • 2005 Hearing Journal survey
    34% use routinely

Most Recent Survey
Hearing Journal, May 2010

• Mueller and Picou
  • N=309 audiologists, 111 HIS
  • Results:
    45% of audiologists routinely use probe mic measurements
    Half of audiologists who have the equipment don’t use it

• But....
• Mueller put in a “lie detector” question
• Fabricated a test: “Binaural Summation Index”
  • Asked if they used it
  • 20% of audiologists said they used it and the test doesn’t exist
• So if there’s a 20% lie factor, then maybe only 45% - 20% = 25% actually do probe mic measurements

Other Recent Voices on the Issue
• Recent Consumer Reports article on hearing aid provision
  • 2/3 of hearing aids not fit properly
  • Probe mic testing is a “must have” procedure for every consumer purchasing hearing aids
• Audiology Today (Sept-Oct, 2009)
  • Catherine Palmer suggests it may be “unethical” not to do probe mic measurements as it is recommended “best practice”
  • Probe mic measurements significantly related to subjective benefit and handicap reduction
  • If probe mic measurements done, users more likely to
    • Recommend hearing aids
    • Recommend the audiologist
    • Repurchase the same HA brand

Why Aren’t More Audiologists Doing Probe Mic Measurements?
• Is it time?
  • Only takes 10-15 minutes
• Is it audiologists don’t understand probe mic techniques?
  • Possible, but it’s not hard to do or understand
• Is the equipment too expensive?
  • Costs $7k - $12k
  • Profit on 5-10 hearing aids
  • My Verifit is 8 years old, so 1 HA/year
If I Were A HA Patient and Was Informed of the Following…

• All professional associations recommended these measures be made, it’s “best practice”
• Everyone agrees speech should be audible and without the measures you don’t know if it is or not
• The measurements only take 10-15 min
• The settings I’ll use 16 hours/day for 5 years were based on simulations and my audiologist didn’t know what my hearing aids were doing in MY ears
• The measurements weren’t made and I paid $5k-6k for the hearing aids…I’d be MAD!!

Maybe So, But…

• You should be doing it
• Doesn’t take much time
• Not expensive
• You are verifying what the hearing aid is supposed to do
• It’s best practice
• Patients deserve it
• “Just do it”
Fitting Options for Unilateral Sensorineural Hearing Loss

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Options

- **Baha**: Implanted to mastoid of poorer ear after exploring other fitting options (Cochlear America; Oticon Medical).

- **Transcranial CROS**: Power BTE or custom hearing aid fit to poor ear.

- **TransEar**: Quasi-BC aid fit to poor ear (Ear Technology).

- **Wireless CROS**: Transmitting microphone on poor ear and receiver in the better ear ( Phonak, Unitron and Interton). 2010-2011 was a year of very exciting advances!

- **Sound-Bite**: BC via teeth. Approved by FDA Jan 2011.
Baha

Cochlear Baha BP100

- 12 channel sound analysis
- Three programs
- WDRC
- Automatic adaptive multi-channel DM
- Automatic noise management
- Fb cancellation
Oeding, Valente and Kerckhoff (2010)

Mean Benefit

<table>
<thead>
<tr>
<th>Condition</th>
<th>OM</th>
<th>DM</th>
<th>Diffuse 180</th>
<th>Closed</th>
<th>Open</th>
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<tr>
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<td>5.7</td>
<td>4.6</td>
<td>3.5</td>
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<td>Environment</td>
<td>2.5</td>
<td>1.1</td>
<td>4.9</td>
<td></td>
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<tr>
<td>Better Ear</td>
<td>6.5</td>
<td>1.6</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Oticon Medical Ponto and Ponto Pro

- Automatic Adaptive DM
- Four memories
- Noise Reduction
- Data-logging
- 10 band frequency shaping
- Self Learning VC
- Linear signal processing

Testing Directional Microphone

Front speaker
Ref mic
Back speaker
TU-1000 Skull simulator
Transcranial CROS (Power aid to poorer ear)
REM to verify transcranial CROS

Probe microphone in “dead” ear and measuring threshold in dB SPL near TM.

Goal to verify that the measured REAR is above TCT.

In this case, REAR$_{70\&80}$ is “above” TCT and assumed to be audible. REAR$_{50}$ is above TCT only at 3000-4000 Hz.
Measuring TCT

- Insert Earphone from Audiometer
- Probe Microphone

Frye 7000

- Microphone Calibration
  - Coupler Microphone
  - Reference Microphone
  - Probe Microphone 51.1 dB
Goal to verify that the measured REAR is above TCT.

In this case, REAR_{70 \text{ & } 80} is "above" TCT and assumed to be audible. REAR_{50} is above TCT only at 3000-4000 Hz.
BC Vibrator
Second bend

Not acceptable (too short)

Great impression!

TransEar

TransEar target area

ear canal
Output Response of New TransEar Vibrator
Programmability of TransEar

- Adjustments via stand-alone NOAH module:
  - Overall Gain and output
  - 12 “handles” between 200-7200 Hz to adjust frequency response
  - Up to 4 memories
  - Compression Threshold (40-70) and Compression ratio (1:1 to 4:1) in four channels.
  - Signal processing (WDRC; AGC-O; Linear)
  - Noise reduction (off; 7, 10, 13)

Sound-Bite
Approved by FDA in Jan 2011
Sound-Bite Hearing System (Sonitus)

- Non-surgical
- Dental retainer containing the battery and actuator
- DSP BTE with a microphone in the ear canal
- Deliver signals >12,000 Hz.
- Rechargeable batteries for BTE and retainer

Mic
DSP BTE w/wireless signal to retainer

Actuator and FM receiver

Retainer worn in upper molars w/wireless pickup to actuator

Popelka (2011): Sound-Bite

BC transducer + electronics + FM receiver

Rechargeable Battery

Hermetically sealed
Popelka (2010): Sound-Bite Insertion of Mouthpiece

Sound-Bite Procedures

• Patient has appointment with ENT
• Refer to partner dentist for exam + impression for SoundBite
• Refer back to dentist for physical fit of device
• Refer to audiologist for the fitting
  • No current information on programming or verification
CROS

First.... Concern with CROS
Example of Good **Unaided** Listening in a Patient With SSD

Unaided

This Can Easily Convert to Poor **Aided** Listening

Aided
This situation could be improved if the transmitter side had:

a. VC so the patient could reduce gain (or turn off) if noise is present on that side…and

b. Effective NR that would attenuate, rather than amplify if an unmodulated signal (noise) was detected and amplify only if a modulated signal (speech) was detected.

Now, an Example of Poor Unaided Listening

Speech Noise

Unaided

Normal Hearing

Unaided
A Typical CROS Candidate

a. Sound is picked up by microphone on the unaidable (left) side and transferred to an ear with normal or near normal hearing (right).
b. Open earmold on the “good” side.
c. Need some HF HL
Two Recent Advances with CROS/BICROS Hearing Aids

New Wireless System from Phonak

Receiver side has active DM in CROS/BICROS Mode
Tandem 16: NR on Receiver Side

Green: No NR  
Pink: Min NR  
Blue: Max NR

Overall decrease of 8 dB

Tandem 16: NR on Transmitter Side

Green: No NR  
Pink: Min NR  
Blue: Max NR

Overall decrease of 6 dB
The next step is releasing a CROS/BICROS system with NR AND DM as is currently available to all other hearing impaired patients.

Thank You
How Did I Get Here?
Vestibular and Balance Disorders Niche OR What I Did Last Summer

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Lets Start at the Beginning

• I was taken from a simple reed basket floating down the Nile River.
So, What is My Niche?

• While involved in developing testing protocols and basic interpretations – administering the tests is not what I do regularly.

• I perform a detailed office visit that consists of:
  • Detailed neurotologic history
  • Complete past medical history
  • Full systems review
  • Direct ENT, Vestibular, balance and neurological examination
  • Integrate and interpret the direct office and laboratory studies in the context of the history to arrive at a most likely diagnosis and recommendations for treatments / and or further work up.

So, What is My Niche?

• My clinical activities fall in the area of a physician extender program.

• Each patient I see is at the request of a physician caring for the patient or standing request before being seen by the physician of record. Therefore, my services to the patient are at the request of the physician of record.

• In this situation my activity does not constitute practicing medicine without a license.
How Did This Develop? Is There a Need for Such?

- The majority of dizzy patients are referred to ENT
- An ENT practice survives financially in the OR not in the clinic – therefore seeing a large number of patients in the clinic is what filters patients for the OR
- To properly evaluate, diagnosis, develop and explain a plan to a chronic dizzy patient requires a 1 hour appointment not the typical duration for ENT

How Did This Develop? Is There a Need for Such?

- Ford Hospital 1983-1987 concept was developed while I was administering and interpreting laboratory testing
- 1987 – 1995 with the help of Dr. Telian at the U of MI we further developed and revised the concept of a physician extender program
- Further modifications then occurred at U of PA in the context of a truly integrated balance and dizziness center. This brings us to the present formulation of the services I provide and teach
Knowledge & Skill Set

• Thorough basic understanding of A&P not just of the auditory and vestibular systems but of the body as a whole and the interaction among the systems
• Detailed & advanced understanding of the auditory and vestibular systems both peripheral and central including the peripheral and central ocular motor system
• Detailed understanding of the mechanisms for balance and gait and the relationship to the vestibular system

Knowledge & Skill Set

• A strong understanding of the discipline of ENT in general and detailed understanding of the medical and surgical aspects of neurotology
• A strong understanding of the neurological and psychological disorders that effect balance and can cause dizziness
• A thorough understanding of the pathophysiology and medical, surgical and physical therapy treatment of dizziness and balance disorders and the various disorders that produce symptoms in this arena
Development of the Knowledge & Skills

- My goal was under the premise that the inner ear does not function in isolation – therefore you need to know about the other systems and the relationship – This was done with:
  - Taking the didactic courses from the Medical school at U of IA
  - 3 month intensive U of IA ENT resident orientation including 60 hours of head and neck dissection
  - Attendance at ENT ground rounds
  - Observation and working with Neurology, ENT in both clinic and surgery

Can You Get Paid?

- Originally by “incident to” with the Physician
- Cash
- Use of E&M codes
  - Audiologists can not use E&M for any CMS insurance
  - Can use E&M for other insurance carriers – in general
  - Combination of Cash (ABN – waver) for CMS and E&M to others
Is This Niche Useful?

- It adds an additional layer of complexity with scheduling and additional charges for the patient / insurance.
- It allows the physician involved to see the patients in a shorter time frame and with an advanced suggestion of diagnosis.
- It gives those struggling with a complex dizzy patient an alternative avenue for suggestions.
- Anecdotally – yes it is of use – no formal studies – real proof would be does it improve the level of care for the patients?

The End of My Summer Report
Holding Humanitarian Hearing Healthcare Providers to High Standards

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Common Threads
Humanitarian Priorities??

- Where are those Free Hearing Aids?
- Who will donate supplies?
- How can I find funding?
Primary Objective: Sustainability

Effective Programs

Coupled With . . .

ACCEPTABLE & ACCOUNTABLE behavior
“What Happens in Africa, Stays in Africa”

• Who SNORES
  • Who belongs to THOSE undergarments
    • Hygiene
    • Sleep talkers confessions
      • Frustrations shared

Guidelines – What is Acceptable

• Legally

• Ethically

• Professionally
Ethical Principles

Standards by which the profession and the individual members determine the propriety of conduct. Assures:

Professional Integrity
Public confidence in the integrity of the services provided
Humanitarian?
Code of Ethics: Conduct and Relationship with Patient
(ISA – Code)

a. Continuing Education is in the best interest of the profession
b. Referral shall utilize all resources available
c. shall accept and seek full responsibility for the exercise of judgment within, but not limited to, the areas of his/her expertise.
• Shall not guarantee outstanding results
• Shall exercise caution not to mislead persons to expect results that cannot be predicted.
d. Shall hold in professional confidence all information and professional records concerning a patient.

e. Conduct in Regard to Colleagues.
• Shall avoid disparaging, pejorative, and/or inaccurate remarks or comments about professional colleagues
• Shall conduct himself/herself at all times in a manner which will enhance the status of the profession.
• Shall be supportive to individuals and organizations with whom he/she is associated.
• Shall not agree to practice under terms or conditions impair the proper exercise of professional judgment and skill

f. Maintenance of Records of services provided and observe all laws or rules and regulations
g. Shall not participate with other health professionals or any other person in agreements to divide fees or to cause financial or other exploitation when rendering his/her professional services.
h. Shall not delay furnishing care to patients served professionally, without just cause.
i. Shall not discontinue services to patients without providing reasonable notice of withdrawal
j. Practice good safety and sanitation procedures
Professional – Ethical Dilemmas?

1. Minister of Health happy to welcome your teams immediate provision of 1000 – 4000 instruments (cochlear implant or hearing aids) with little to no hearing health professionals in the country.

2. OB-GYN personally provided vision screening last year in Nigalal, now he’s wanting to personally offer hearing screenings in his mission this year.
1. Minister of Health happy to welcome provision of instruments (cochlear implant or hearing aids) with little to no hearing health professionals in the country.

2. OB-GYN personally provided vision screening last year in Nigeria, now he’s wanting to personally offer hearing screenings in his mission.

3. You hear about a cohort of deaf children at an orphanage with no deaf education available in the province, what would be the communication choice you will advocate in your mission: C.I., hearing aids, or manual?

4. A very enthused professor who has never left the U.S. borders has been enlisted by one of her patients to join him in an existing mission of providing mobility options to local handicapped in Katmandu. But, the desire of this professor is to offer newborn hearing screening at one of the local hospitals, once she arrives.
“Big ideas are important, enduring, and transferable beyond the scope of a particular unit”  
(Peace Corps)

Providing an Ethical Program

• What is the purpose of your program?  
  Categories of service
Levels of Health Service

• The Community
• The Community Health/Promoter
• Primary Care Nurse Practitioner/Clinical Assistant
• Primary Care Health Workers with Specialty Training/skills

Providing an Ethical Program

• Purpose: categories of service
• Seeking Guidelines
So . . . Structures and Guidance Available for Humanitarians?

Local Health Ministry
World Health Organization
International Society of Audiology

Disabling Hearing Impairment (degrees)
Adults: 41 dB or greater
Children (<15 years): 31 dB or greater

In Better Ear
Considered Permanent
Unaided thresholds @ 500, 1000, 2000, 4000 Hz
Providing an Ethical Program

• Purpose: categories of service
• Guidelines
• Strategies & Structures
  • Test or Screen
  • Recording results
  • Maintaining records
  • Follow-up
**Documentation Strategies**

**Networking Opportunities**

- **Coalition for Global Hearing Health**
  (http://coalitionforglobalhearinghealth.org)
- **Tele-audiology Network**
  (http://www.teleaudiology.org)
- **ANNUAL Lunch time meeting of humanitarian audiologists at AAA-Chicago on Friday, 8 – April (BYOL)**
- **Large Email distribution list of humanitarians world-wide**
http://www.isa-audiology.org/human.html
Coming 2011:

**Humanitarian Hearing Health: Planning for the Best Effect**

Editors:
J.L. Clark & J. Saunders

Auban, Inc. Publishers

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**BOOKS**

**Upcoming Workshops 2011**

- AAA Webinar: 16-March 2011 (1:00 p.m. CST)
- AAA Chicago – Featured Session: 9-Apr 2011
- Global Coalition for Hearing Health Conference – full day TBA
Humanitarian Priorities??

✓ Where are those Free Hearing Aids?
✓ Who will donate supplies?
✓ How can I find funding?

WHEN IN DOUBT - Humanitarians hold themselves to the same standard they practice at home.

PROFESSIONAL ORGANIZATIONS become involved in Humanitarian Efforts by directing their members to following established guidelines and resources.
The World NEEDS Our BEST
A Step Beyond
An Audiologist in Aerospace Medicine Research

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How Did This Happen?

• Learning is challenging fun
• There is no market on knowledge
• Passion underlies invention
• Everything you understand is easy
Lesson Learned

- Went to Australia in 1976 to complete my clinical fellowship
- Scientist at U. Melbourne thought you could actually implant someone and restore hearing
  - Many skeptics including myself
  - It impacted my attitude on what was possible

History Repeats Itself

- 1980 developed a newborn hearing screening program using ABR
- For 5 years assisted with ABR intra-operative monitoring for pediatric brain stem tumors
- Protocols developed for hearing and neurological assessments, including evaluations in determination of brain death
Over and Over Again

- Identified link between severe to profound hearing loss and bronchopulmonary dysplasia
- Newborns given the drugs gentamicin and furosemide concurrently
- Research in Food Science and Nutrition to evaluate magnesium deficiency as a factor
- Today conduct protocols looking at magnesium deficiency in hearing loss and tinnitus

From Pediatrics to Adults

- In 1987 joined Mayo Clinic
- Predominately an adult population
- No longer performed intraoperative AEPs
- Practiced every area of audiology which broadened my clinical knowledge and skills
Knowledge Knows No Limits

- Past experience put to use in new question
- Attitude of “can do”
- Discovery is rewarding and fun
- Team becomes infectious with wonder
- New ideas have tremendous momentum

One Good Turn Deserves Another

- DOD asked to create the illusion of 360 degree rotation in fixed seated position
- Helicopter pilots needed vestibular input for simulation of rotor failure during simulator training
- How do we do that?
- Do we want to do that?
History Repeats Itself at Mayo

• Hypoxia from high altitude
  • Oxygen mask
  • Bailout bottle

• Acceleration forces
  • G-suit
  • Anti-G straining maneuver
AMVRL Areas of Research

- Spatial disorientation
- Hypoxia
- Acceleration
Transition From Clinic to Laboratory

- Clinical background
- Aerospace
- Bioengineering
- Bridge gaps with reading and discussion
- Testing using all vestibular tests for initial Phase I funding

Unique Environment at Mayo

- Collaboration
- Equipment
- Team of clinicians
- Research process
- Sufficient time to conduct study
Somatogyral Illusion

• What do we know about the underlying physiology?
• Can we re-create with artificial stimulation using small amount of electrical current?

The Answer is “Yes”

• Galvanic vestibular stimulation
  • History
  • Physiology
  • Effects on the vestibular system and balance

Video clip from the accompanying DVD for *Balance Function Assessment and Management* by G. P. Jacobson and N. T. Shepard (Eds.). Copyright © 2008 Plural Publishing, Inc. All rights reserved. Used with permission.
GVS Induced Nystagmus

Right Warm Caloric with GVS Suppression
Yaw, This is True

- With multiple electrodes can have activation pattern of GVS induced perception in all three planes of rotation, including yaw, pitch, and roll.

http://exploration.grc.nasa.gov/education/rocket/rotations.html

The Answer is “Yes”

Now you can create illusion by using galvanic vestibular stimulation but how do you quantify perception?
• Recording perception of vestibular illusion
• Developed a dose response for level of current and perception
• Findings are now integrated into flight simulation to create a virtual head movement coupled with moving visual field
Avatar Recordings of GVS Induced Perception

- Applied GVS dose response synchronized to moving visual field in helicopter flight simulator

Mitigation of Simulator Sickness

- Applied GVS dose response synchronized to moving visual field in helicopter flight simulator
Vestibular Cueing

- AeroStim chair
- Quantify the effects of GVS in the mitigation of perception during movements in pitch and yaw
- Limitations of rotary chair for flight simulation

Pitch This Idea
AeroStim Chair

Scope of AMVRL
25 Grants in Past 6 Years

- Hypoxia and balance
- Hypoxia and eye tracking
- Anti-G straining maneuver
- Mitigation of simulator sickness
- Multi-sensor simulation suit
- Coupling of visual movement with virtual head movement using GVS
- Mitigation of motion sickness
- Vestibular cueing
Simulated Mars Environment Using Holodeck

Build a Foundation of Knowledge and Experience

- Gather and record data
- Videotape everything
- Organize
  - Build a team
  - Audiology
  - Neurology
  - ENT
  - Research support
- Intellectual Property
Clinical Integration

- Mitigation of vertigo in Meniere’s
- Mitigation of motion sickness
- Vestibular cueing in balance disorders
- Hypoxia in balance disorders
- GVS as an assessment tool

Criteria for Results

- Intellectual curiosity
- Passion to accomplish one’s goals
- Be a good team member
- Failure is one step to success
- The process itself is a reward
- Build relationships
Transition in career
Managing Interesting Pediatric Cases

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Case Example 1

• 14 month old female
  • Hospitalized for meningitis
  • Parents concerned about hearing
Case Example 1

- Inpatient Audiology Consult
  - Type C tympanograms bilaterally
  - Absent TEOAEs bilaterally
  - ABR
    Absent neural responses at the limit of the equipment (90 dB nHL) bilaterally

CT scan ordered
- No evidence of ossification
- Serial scans
Case Example 1

- Initial Outpatient Audiology Consult

- Hearing Aid Fitting
  - 8 days following ABR
  - Right loaner power BTE hearing aid
  - SAT = 45 dB HL
Case Example 1

Spontaneous recovery of hearing in right ear approximately 3 months post Meningitis

- Parents report
  - Over 50 words at 20 months
  - 115 words at 23 months
Case Example 1

High frequency hearing declines

- Preschool behavior concerns
  - Lack of participation in group activities
  - Aggressive towards peers
  - Difficultly maintaining attention
Case Example 1

- Parents concerned with speech and language
- Speech and language consult
  - Receptive and expressive language skills are falling in the low average to mildly delayed range for age
  - Speech production skills are significantly delayed for age

Case Example 1

- Cochlear implant evaluation
  - Difficulty in noisy environments
  - Difficulty with multiple speakers
  - 1st indication of slight ossification
  - Aided speech perception testing
    - 33% MLNT easy
    - 16% LNT easy
FDA Guidelines: Pediatric Cochlear Implantation

• Ages: 25 months to 17 years, 11 months
  • Severe-to-profound sensorineural hearing loss in both ears
  • MLNT scores of 30% or less in best-aided condition
  • Lack of progress in the development of auditory skills
  • No medical contraindications
  • High motivation and appropriate expectations

Case Example 1

• First week of implant use
  • Full time use
  • Alerted parents if processor wasn’t working
• One month follow-up
  • Behavioral changes at school
  • Observations at home
Case Example 1

• 2 months of CI use
  • Parents report “clearer speech”

Case Example 1

• Speech & lang. at 5 months CI use
  • Made gains in receptive skills that exceed the rate of maturation
  • Very socially engaged and willing to work on communication skills
  • Speech-production skills remain significantly delayed for age, with progress occurring at about the rate of maturation
Case Example 1

- 5 months post initial activation
  - Left cochlear implant only
  - 70% GASP sentences
  - 60% GASP words
  - 42% MLNT easy
Case Example 1

- 46 months post initial activation
  - Left cochlear implant only: MLNT Easy: 92% words correct
  - Right hearing aid only: MLNT Easy: 92% words correct
  - Bimodal: CNC: 86% words correct
    AzBio-C = 100%
    BKB-SIN = 5 dB SNR

Case Example 2

- 10 year-old male
  - Progressive difficulty hearing
  - Listening becomes more difficult when asked to multi-task
  - Increased difficulty listening in noise
  - Dx’d and treated for ADHD
  - Difficulty with handwriting
  - “Not graceful”
  - Tingling in legs and feet
  - Abnormal gait
Case Example 2

• Background/medical history
  • Passed newborn hearing screening
  • Hyperbilirubinemia
  • Recurrent eustachian tube dysfunction and PE tube placement until age 5

• School Report
  • 1st grade - passed hearing screening
  • 3rd grade – passed after a re-screen of the left ear at 2000 Hz
  • 5th grade – failed at 500 Hz in both ears and 2000 Hz in left ear
  • Teachers have concerns regarding hearing
Case Example 2

- Otology consult
  - M.D. observed difficulty in office
  - MRI ordered
    - No evidence of cochlear malformations or other concerns
  - Neurology consult ordered

- Medical work-up
  - Genetics
    - Charcot-Marie-Tooth syndrome
  - Neurology
    - Sensorimotor peripheral neuropathy
  - OT and PT
    - Gait disorder
  - Ophthalmology
    - Ruled out any ocular concerns
Case Example 2

Initial Hearing Evaluations

- SRT = 25 dB HTL
- Word Recognition:
  - right = 93% correct
  - left = 35% correct
- Normal tympanometry

- Distortion Product Otoacoustic Emissions (DPOAEs)
  - Present 750-8000Hz bilaterally
- ABR – Click stimulus 90 dB nHL
  - Left - Absent neural response
  - Right – Delayed latency & poor morphology of wave V
  - Cochlear microphonics present bilaterally
Case Example 2

- Diagnosed with Auditory Neuropathy Spectrum Disorder (ANSD)
  - Normal outer hair cell function
  - Absent or abnormal auditory nerve function

- ANSD
  - Impaired temporal processing
    - Delayed gap detection
    - Delayed detection of amplitude modulation
    - Impaired binaural processing
  - Difficulty with speech perception in noise
Case Example 2

- Speech Perception Testing
  - Binaural:
    CNC Monosyllabic Words 80% correct
    AZBio-C Sentences in quiet 100% correct
    BKB SIN 10.25 dB SNR
  - Right:
    CNC Monosyllabic Words 90% correct
    AZBio-C Sentences in quiet 99% correct
    BKB SIN 10.5 dB SNR
  - Left:
    CNC Monosyllabic Words 72% correct
    AZBio-C Sentences in quiet 99% correct
    BKB SIN 9.25 dB SNR

Recommendations:
- FM system trial in school (iSense) – two thumbs up and says “it’s working great.”
- Follow annually
Case Example 3

- Passed NBHS via OAEs
- Speech/language delays
- Hearing Loss identified at 20 months of age at outside clinic
- Inconsistent responses to sound
- Absent ABR waveforms reported by outside clinic

First ear specific behavioral audiogram at 21 months
Case Example 3

- Fit with loaner bank hearing aids
- REM showed hearing aids met DSL i/o targets for audibility of speech (60 dB SPL)
- Received weekly SLP services
- Primary mode of communication through sign language

Mayo SLP evaluation at 22 months
- Ski*Hi Language Development Scale revealed receptive/expressive language of 14-16 months
- Preschool Language Scale revealed receptive/expressive language of 12 months
- Stark Assessment of Early Vocal Development (SAEVD-R) revealed expressive skills of 3-8 months
Case Example 3
DPOAEs at 23 Months

Case Example 3
ABR at 23 Months
Case Example 3
ABR at 23 Months

- CT scan at 23 months
  - The vestibular aqueducts are normal in caliber
  - The bony labyrinths and ossicles are normal in appearance bilaterally
- Otologist reviewed SLP evaluation, CT scan, ABR, & OAE results & ordered a CI evaluation
Case Example 3

LittlEars questionnaire

• No progress in auditory skills after 3 month hearing aid trial

• Auditory skills are equivalent to those of children with normal hearing at 8 or 9 months old

• She remains significantly delayed in her auditory skills even with HAs and is judged to be a cochlear implant candidate
Case Example 3
SLP Follow-up at 25 Months

- Has been wearing HAs for 5 months now
  - Ski*Hi Language Development Scale continues to exhibit expressive/receptive skills at roughly a 14 to 16 month level
  - Receptive/Expressive Emergent Language Scale (REEL-3) continues to be at a level of less than 1 year
  - Preschool Language Scale (PLS-4) continues to be at a level of less than 1 year
  - Rating of vocalizations using the Stark Assessment of Early Vocal Development (SAEVD-R) continues to place her at around a 3-8 month level

Case Example 3

- Cochlear implantation of right ear at 25 months
- Initial activation at 26 months
- LittleEars Auditory questionnaire at 3 months post initial activation revealed 32/35 points
Case Example 3
SLP Follow-up

• 6 months post initial activation
  • Preschool Language Scale (PLS-4) improved from 12 to 21 months

Case Example 3
6 Months Post Activation

Audiogram
Case Example 3
9 Months Post Activation
2 Years, 11 Months

Testing at 60 dB SPL
• GASP=100%
• MLNT Easy
  58% words
  85% phonemes
LittEars: 35/35

PLS-4: normal

1 Week Later.....

• Baby sister is born (Case Example 4)
• She does not pass NBHS via AABR
Case Example 3
1 Month of Age

Diagnostic testing
• Present OAEs
• ABR consistent with ANSD

6 Months Later

Case Example 3
(3 yrs., 5 mo.)
1 year, 5 months post implant @ 60 dB SPL
• MLNT Easy: 100%
• LNT Easy: 80% words, 82% phonemes

Case Example 4
(7 months of age)
• LittlEars: 5
• Parents do not want to go through a HA trial
1 Month Later….
Case Example 4

- MRI at 8 months
  - Intact cochlear nerves bilaterally
  - The brain and inner ear structures are morphologically normal

Case Example 4
ABR at 8 Months
Another 2 Months Later…

Case Example 3
- Receives her 2nd implant in the left ear at 3 yrs, 9 mo.

Case Example 4
- Receives simultaneous bilateral implants at 10 mo. of age

Update

Case Example 3
- 3 month post activation of 2nd implant (left ear)
- Now 4 yrs, 1 mo.

Case Example 4
- 3 month post activation of simultaneous bilateral implants
- Now 13 mo.
Questions?