Pediatric Grand Rounds: LEND Pediatric Audiology Training Program Webinar Series

About the Webinar Series

Funded through the Maternal and Child Health Bureau (MCHB), the MCHB/AUCD LEND Pediatric Audiology Training Program supports 10 LEND programs to increase the didactic content and clinical experience of trainees in pediatric audiology.

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Pediatric Grand Rounds

Webinar Objectives:

Describe challenging pediatric audiology case studies in view of cultural differences

Explain the diagnostic process for arriving at a hearing loss diagnosis

Describe the audiological management provided to children with hearing loss in light of other medical or social challenges

Identify LEND competencies as they relate to the cases presented

Illustrate how LEND core competencies impact care and management of children with hearing loss

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Pediatric Grand Rounds
Agenda

- Welcome: Maternal and Child Health Bureau, HRSA

  Irene Forsman, M.S., RN
- Presentations: Children's Hospital of Pittsburgh of UPMC

  Introduction and moderation by Diane Sabo, PhD
- Presentations: Boston Children's Hospital

  Introduction and moderation by Brian Fligor, ScD
- Q&A

  Submit questions via the chat box
- Evaluation Survey
  - Please complete our short survey to give us feedback

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Diane Sabo, Ph.D.

Jessica Wells

William Dillon

Tessa Utz

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Cochlear Implantation in the Amish Community: A Case Study

William Dillon

Jessica Wells

LEND Trainees
Children's Hospital of Pittsburgh of UPMC
University of Pittsburgh

LEND Competencies

- Cultural Competency
- Conflict Resolution
- Family-Centered Care
- Working with Communities and Systems

Amish Culture

- **Christian religious group**
- **Amish vs. Amish Mennonite**
  - Resistance to technology
- **Populations/Regions**
- **Marriage and forming families**
  - High prevalence for genetic disorders
  - Childbirth considerations

Genetics and Hearing Loss

- **"Founder Effect"**
  - Autosomal recessive disorders
- **Prevalence**
  - 1/250 to 1/500 births (Morton et al., 2003)
- **Related Auditory Disorders/Syndromes**

History

- AH, 6yr female in the Amish Community
- Significant family history of hearing loss and development al disorders
- Identified at birth with bilateral profound hearing loss
- Underwent cochlear implant evaluation and was scheduled for surgery in 2008
- Post-poned surgery until 2012 due to medical issues

Cochlear Implant

- Advanced Bionics HiRes 90K cochlear implant, with Harmony speech processor in the RE
- Consistent CI user, takes off when she is tired or wants attention
- Able to program by using NRI and AH's facial reactions to sounds
- Family's goal is for oral communication
- AH does have her own medical insurance

Take Home Points

- **Can't make assumptions based on Cultural stereotypes**
  - Have to make families aware of every option available to them
- **Be aware of accommodations you may need to make for the family**

Managing Neural Hearing Loss

Tessa Utz

LEND Trainee

4th year Audiology extern:

Children's Hospital of Pittsburgh of UPMC

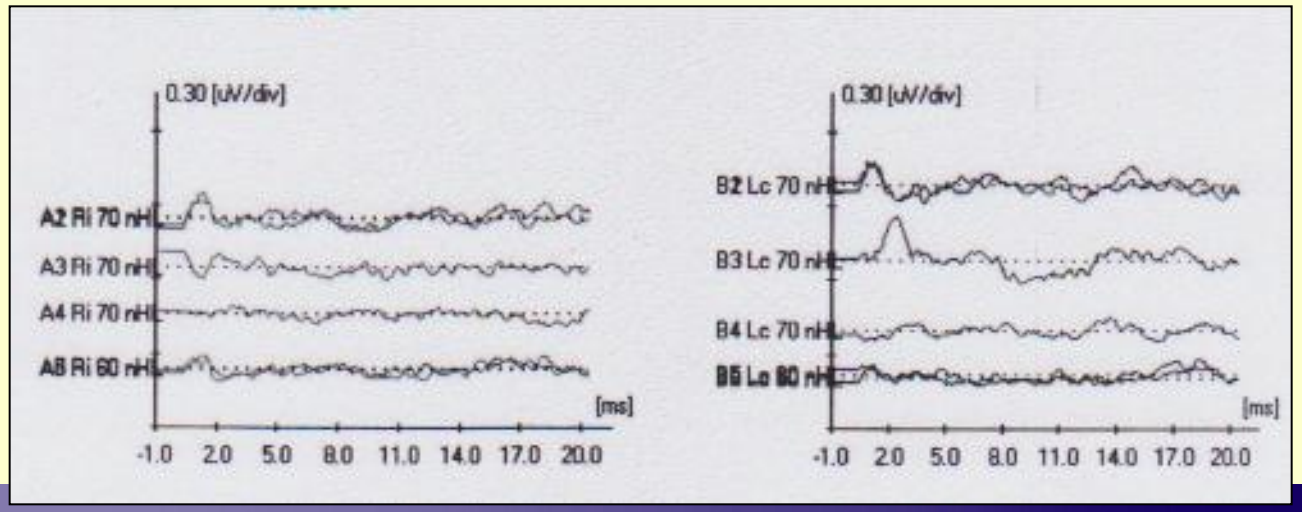
University of Pittsburgh

Objectives

- **Review case history**
  - Diagnosis of a neural hearing loss
- **Discuss reasoning for the current management plan**
- **List the unique challenges of this case**
- **Illustrate how the challenges were addressed**
  - LEND competencies

Case History

- **Diagnosed with kernicterus**
- **Referred to audiology**
  - Neural hearing loss
    - Present OAEs bilaterally
      - Intact sensory function
    - Abnormal ABR bilaterally
      - Disrupted neural function



Case History

	6/14/12	7/18/12	8/14/12	9/13/12	10/24/12
Behavioral Findings	Moderately elevated responses to speech in SF	Mildly elevated responses to speech bilaterally	Normal responses to speech and 2kHz stimuli bilaterally	Inconsistent responses; Possible SAT at 30 dB in SF	SAT obtained at 30 dB in SF
Parental Report	Inconsistent responses to sounds at home	Responsive to voices without visual cues	More consistent auditory responses and variegated babbling	Able to discriminate voices	Consistent responses in quiet; Inconsistent responses in noise

Management

- **Watchful, waiting approach**
  - Need for reliable thresholds
- **Referral to speech language pathology**
  - Track gains in speech and language skills
- **Cochlear implantation?**
  - Parent expectations
  - Recommendations from outside facilities

Challenges

- **Using a waiting approach created stress**
  - Need to respect parental goals while defending management decisions
- **Aspects of case exceeded my knowledge**
  - Need for collaboration with other disciplines
- **Involving professionals from outside facilities was difficult**
  - Need for consistent recommendations
  - Communication with other professionals

LEND Competencies

- **Family Centered Care**
  - Parents as decision-makers
- **Communication**
  - Reduces stress on family
- **Ethics and Professionalism**
  - Acknowledge my own limitations
- **Negotiation**
  - Development of trust to facilitate compromise
- **Interdisciplinary Team Building**
  - The expertise of others is viewed as essential

Conclusion

- **Neural hearing loss can be challenging to manage**
  - Takes time to establish behavioral thresholds and track speech and language development
- **Following LEND competencies can ease the management process**
  - Inclusion of other professionals
  - Acknowledgement of parental goals
  - Development of trust to arrive at compromise

Brian J. Fligor, Sc.D.

Nicole Corbin

Rebekah Tozer

Lauren Shastany

AUCD

4th Year Externship and LEND Fellowship at Boston Children's Hospital

Brian J. Fligor, Sc.D.
Director of Diagnostic Audiology, Boston Children's Hospital
Instructor, Otology and Laryngology, Harvard Medical School

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LEND at BCH, Institute for Community Inclusion

- Audiology: consecutive LEND Fellow 17 years
- Historically one fellow, then two, now three
- Competitively interview across the country
- 25 applicants for May 2012-May 2013
- Based in Diagnostic Audiology, rotate through Cochlear Implant and Pediatric Balance Center
  - Full service diagnostic audiology (>20,000 visits/yr)
  - Musicians' Hearing Program
  - Sound Start™ hearing aid loan program

Face to Face with Cultural Differences

Nicole Corbin, B.A.
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Diagnostic Audiology Program, Boston Children's Hospital,
LEND, Institute for Community Inclusion, Boston Children's Hospital

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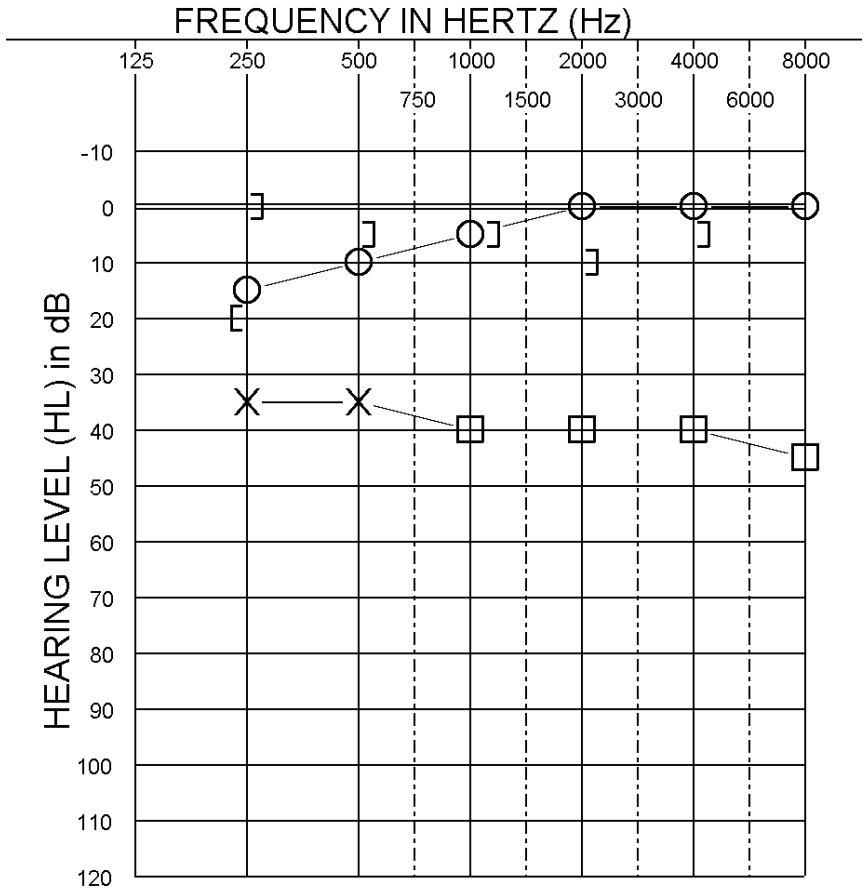
History

- Female, five years of age
- Chinese descent
- Attends school
- Known left stapes displacement
  - Diagnosed at three years of age at outside facility
- August 2011
  - Initial visit to Boston Children's Hospital
    - Four years of age
    - Accompanied by father
  - Asymptomatic, normal speech and language development
    - Possible middle ear exploration and intervention in the future
- February 2012
  - Amplification recommended

Initial Interaction, August 2012

- Father present
- Interpreter of Cantonese present
  - Miscommunication
- Cultural differences emerge



Second Interaction, September 2012

- Mother and younger brother present
- Interpreter of Cantonese present
- Verification of amplification
- Counseling
  - Informational versus adjustment
    - Stepping outside of comfort zone
  - Cultural considerations
  - Realistic expectations

Impressions and Recommendations

Overall Impressions

- Cultural influences
  - Interaction with mother versus father
  - Beliefs surrounding disability
- Communication through interpreter

Recommendations and Conclusions

- It's not about the clinician
  - Put aside own perspectives and world view
  - Initial reaction to father's decision regarding amplification
- Influence of previous patient interactions
  - Every day is a new day

Unilateral Hearing Loss in Osteogenesis Imperfecta Type III

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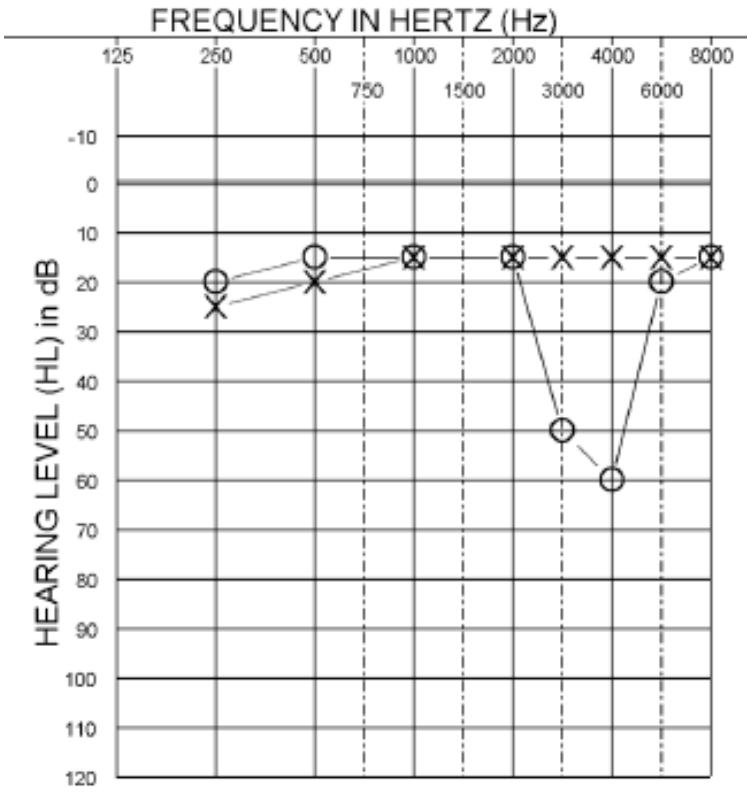
Case History

- 10 year old male
- Osteogenesis Imperfecta type III
- Hydrocephalus and VP shunt on right side
- Followed by multiple clinics at Boston Children's Hospital
- Referred to Audiology by Complex Care Services to assess hearing

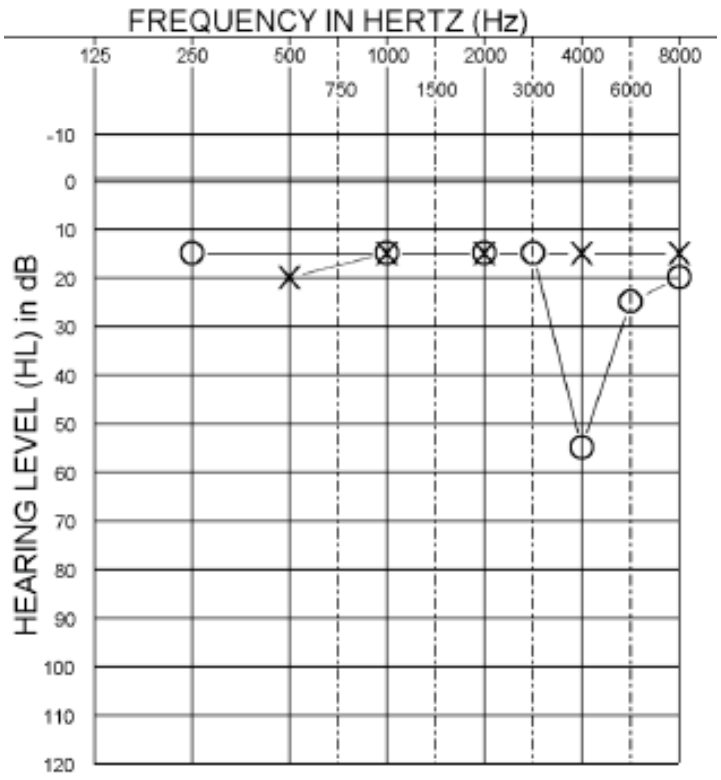
First Visit: July 2012

- Very challenging to test
- Play audiometry used
- Unilateral notch in right ear from 3000-4000Hz
- Could not complete further testing because he became upset and refused to continue
  - refused bone-conduction
- Recommendation: return for further testing to confirm results



Second Visit: Sept 2012

- Immediately accepted wearing headphones
- Conventional test method used
- Could not completed further testing because he became upset
  - Refused bone-conduction
- Confirmed previous test results of unilateral notch in right ear around 4000 Hz
- Recommendation: consult with otolaryngologist regarding hearing loss and return in 3-4 months to further assess hearing status



Impressions and Recommendations

- Re-evaluation in 3-4 months to further assess hearing status
- Consult with otolaryngologist regarding medical implications of hearing loss
- Further recommendations pending re-evaluation and ORL consult

- It is hopeful that by the third visit, he will be more familiar with the task and audiologist that a complete test can be obtained (specifically conductive vs. sensorineural)

References

Spirakis, S.E., & Hurley, R.M. (2003). Unilateral hearing loss in children with shunt-treated hydrocephalus. *Journal of the American Academy of Audiology, 14(9)*, 510-517.

Kaneshiro, N.K. (2011). Osteogenesis imperfecta. In PubMed Health. Retrieved from www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002540/

Osteogenesis Imperfecta Foundation. (2012). Types of OI. Retrieved from www.oif.org/site/PageServer?pagename=AOI_Types

Delayed Onset Sensorineural Hearing Loss with Unclear Etiology

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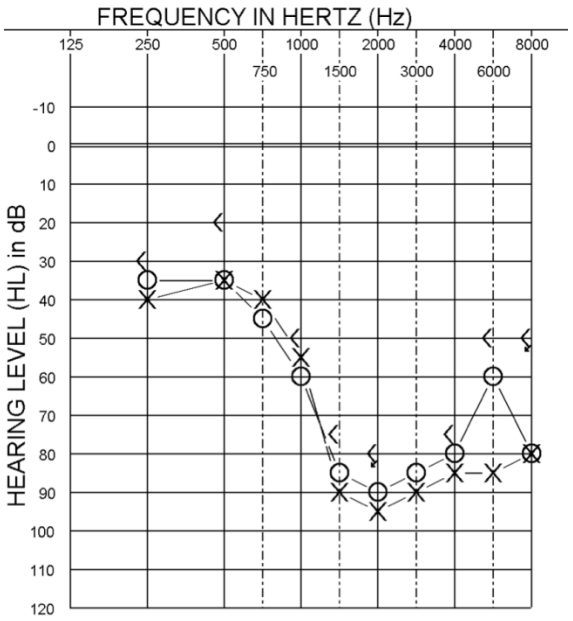
History

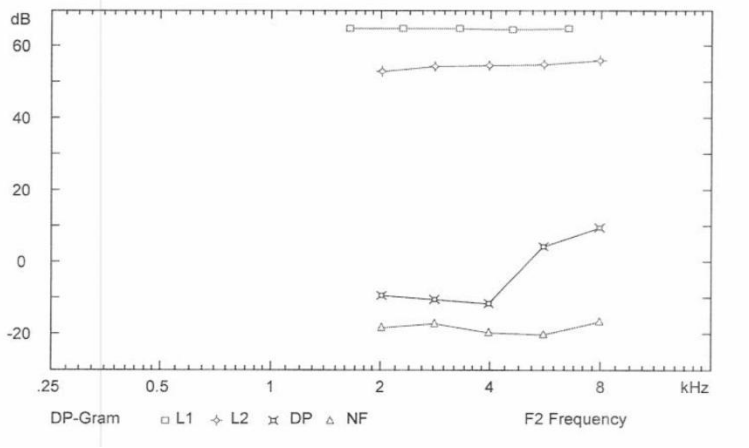
- 4 ½ year old bilingual speaking boy
  - English is a second language
- Referral from pediatrician due to parental concerns
- Passed newborn hearing screening
- Family history
  - Paternal great aunt with hearing loss secondary to a mumps infection
- Typically developing

Audiometric Evaluation

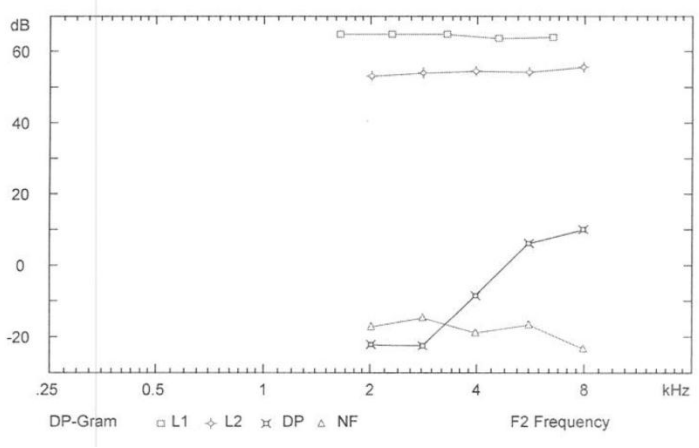


- Bilateral, mild sloping to severe sensorineural hearing loss
  - Reflexes elevated at 500 Hz and absent at 1000 and 2000 Hz bilaterally
- DPOAEs present and robust 6000-8000 Hz

LEFT



RIGHT



Further Testing

- Seen by Otolaryngology
- Evaluation through Deaf and Hard of Hearing Program
  - Speech and Language evaluation
- Fit with binaural hearing aids
  - Poorly tolerated and accepted
  - Reports that he “hears better without the hearing aids”

Differential Diagnosis

- Present DPOAEs at 6000 and 8000 Hz suggesting normal outer hair cell function in that frequency range

- Hearing loss result of MMR vaccine?
  - 28 reported cases of hearing loss following administration of live attenuated measles and/or mumps viral strain-containing vaccines between 1990-2003
  - Hearing loss associated with measles/mumps infection
    - Case report of adult with childhood mumps resulting in bilateral SNHL who had present OAEs but no measurable ABR
    - Temporal bone studies of individuals with hearing loss due to measles/mumps infection show degeneration of the organ of corti

Asatryan et al., (2008); Prieve, B.A., Gorga, M.P., Neely, S.T. (1991); Smith, G.A., Gussn, R. (1976);

Impressions and Recommendations

Next Steps

- Further testing
  - Extended frequency OAEs
  - ABR
  - CI candidacy evaluation?
- Close monitoring of progress with hearing aids
  - Programming changes
  - Validation measures

Final Thoughts

- Cross check principal & critical thinking
- Importance of collaboration & family-centered care

Questions?

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LEND Pediatric Audiology Training Program Webinar Series

JOIN US!

www.aucd.org/audiology

December 13, 2012 (12:00-1:30pm EST):

Featuring: University of Utah Regional LEND, Utah and JFK Partners at University of Colorado, Denver

February 22, 2013 (12:00-1:30pm EST):

Featuring: University of Miami, Mailman Center for Child Development, Florida and the University of South Dakota, Center for Disabilities, South Dakota

March 22, 2013 (11:00-12:30pm EST):

*Featuring: W*aisman Center, University of Wisconsin and Vanderbilt LEND, Tennessee

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Thank you!

Please take a few minutes to complete our survey!

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