Challenges of Audiological Testing with Children with Down Syndrome

Amy Paradis AuD
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CASE STUDY
‘SARAH’

Medical & Birth History

- Full term
- Trisomy 21 – Down Syndrome
- Congenital cardiac defects
- NICU stay of 1 day
- Pacemaker placed at 3 months of age
- Developmentally delayed
- Passed newborn hearing screening in both ears
Audiological Evaluation
6 Months of Age

- **Tympanometry**
  - 1000 Hz: Abnormal both ears

- **Otoacoustic Emissions (OAE)**
  - Absent for both ears.

- **ABR testing** was discussed

- **Recommended**
  - Follow up testing in one month to obtain tympanograms and OAEs.

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Audiological Evaluation
7 Months of Age

- **Tympanometry**:
  - 1000 Hz: Normal for both ears.
  - 226 Hz: Low middle ear compliance in both ears.

- **DPOAEs**:
  - Attempted testing however could not complete due to patient noise and tolerance of the probe tip.

- **Behavioral Audiology**
  - Attempted testing, however patient had poor head and neck control

- **Recommendations**:
  - Attempt behavioral testing in 1-2 months

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Audiological Evaluation

- 6 months: Did not condition. Poor head and neck control
- 7 months: Responded to Speech stimuli
- 9 months: Responded to speech & tonal stimuli
- 10 months: Responded to speech & tonal stimuli (.5-4KHz)
- 13 months: Normal responses in left ear with insert earphone
- 22 months: Responded to speech and tonal stimuli only in soundfield
- 23 months: Responded to speech and tonal stimuli
Tympanometry

9 months 22 months 23 months

Otoacoustic Emissions

9 Months

Right & Left Ear: Present with low amplitude at all frequencies. High patient noise

10 Months of Age

Left Ear Right Ear

Right ear: Cannot interpret - poor probe stability
Left ear: present at one frequency and present with low amplitude at the rest of the frequencies
Tympanometry and OAE Questions

1. Is this type of tympanogram considered to be ‘normal’ for Sarah?

2. Does Sarah have a middle ear pathology that is being missed by the pediatrician or ENT?

3. If Sarah’s ears are clear of fluid, why am I not able to get more robust OAEs?

Audiologic Information

- Stenotic Canals: wax build up, ear canal collapse with headphones
- Otitis Media: PE tubes are common
- Hearing Loss: conductive, sensorineural & mixed
- Multiple Medical Conditions: sedation risk
- Developmental Delay: poor test reliability & possible suprathreshold responses
Down Syndrome

- The most common chromosomal disorder in the world
- Occurs across all races and economic levels

Otitis Media

- Otitis Media may place an additional burden on children as it increases their difficulties in hearing and their language learning.
- Children with Down Syndrome are reported to have a high incidence of recurrent otitis media.
- Middle ear problems account for 83% of hearing loss in children with Down Syndrome. Hearing thresholds were found to be elevated (over 15-20 dB HL) in one or both ears in 76% of the children with Down Syndrome. (Balkany, Mischke, Downs & Jafek, 1979).
- Shott, Joseph & Heithaus (2001)
  - 81% of children with Down Syndrome had abnormal hearing levels
  - 83% of the patients required Pressure Equalization Tube placement due to chronic otitis media with effusion.
  - 2.3% had remaining hearing loss after PE tube placement

Tympanometry & OAEs

- Driscoll et al. (2003)
  - 41.7% of subjects failed tympanometry screening (Type B or C)
  - 81.5% of subjects failed TEOAE screening
- Satwant et al. (2002)
  - 35% of subjects had type B tympanograms
- Hassman et al. (1998)
  - 56% of subjects had Type B or type C tympanograms
  - Low amplitude for DPOAEs
Behavioral Audiologic Testing

- Greenberg (1987) found that VRA is more successful with children with Down Syndrome when they are developmentally 10-12 months of age.
- Greenberg completed the Bayley Scales of Infant Development (BSID) to determine the infant’s mental age equivalent.

What degree of hearing loss do we need to rule out?

Mild Hearing Loss

Did I rule out a Mild Hearing Loss?

Right Ear - OAEs
What is the Impact of Mild Hearing Loss?

- Risk of Educational Delay
- Risk of Speech and Language Delay
- Risk of Social/Emotional difficulties

What is the Impact of a Late Diagnosis?

- Intervention services
  - Implementation of early intervention/school support services
    - Hearing support services
    - Preferential seating in the classroom
  - Potential delay in fitting of amplification/FM

Things to think about….

- Are tympanometry and OAEs the best way to evaluate a young child with Down Syndrome?
- Can we interpret tympanometry and OAE test results the same as a typically developing child?
- Is there a way to rule out a mild degree of hearing loss in the Down Syndrome population at a younger age?
- How does a mild hearing loss affect a child with Down Syndrome?
- What is the average age of diagnosis of hearing loss in this population?
Thank you!

Amy Paradis AuD
The Children’s Hospital of Philadelphia
paradisa@email.chop.edu
267-426-6837

Resources


Pediatric Grand Rounds:
Challenges of Diagnosis & Management of Fluctuating Hearing Loss
Melissa L. Caine, AuD
Pediatric Audiologist
Case Overview

- 15 year old male
- Fluctuating hearing loss of unknown etiology
- Complex medical history
- Diagnostic test battery
- Management plan

“Ethan”

- First visit with CHOP audiology in fall of 2006 in conjunction with otolaryngology – 11 years old
- Testing was consistent with:
  - Patent PE tubes bilaterally
  - Slightly elevated SRT with good discrimination
  - Absent otoacoustic emissions
  - Moderate rising to mild hearing loss
    - Elevated bone conduction
    - Reliability concerns

Audiologic Results: 8/31/06
Patient Follow-up

- Ethan became a case of lost to follow-up
- Reappeared in ENT/Audiology in July 2008 – 13 years old
- Chief compliant was difficulty hearing and increased communication issues

Audiologic Results: 7/2/08
Observations

- Patient present with mother and father
- He was able to answer “yes/no” questions
- Limited eye contact and conversation
  - Not age appropriate
- Articulation errors
- Needed encouragement and reassurance during behavioral testing
  - Parents not present in testing booth

Questions & Concerns

- Significant change in hearing loss
  - Sudden or gradual
- Pseudohypascusis vs. threshold shift?
- When is it appropriate to introduce amplification?
- How to perform effective hearing aid evaluation?

What Else Is Going On?

- Craniopharyngioma with resection 2002
- Borderline prolonged QT syndrome
- Developmental delay secondary to perioperative stroke
- Panhypopituitarism
- Visual field deficits
What Else Is Going On?

- Hypothalamic syndrome
- Obesity
- Obsessive compulsive tendencies
- Conversion Disorder
- Immunoglobulin deficiency
- Learning disability
  - One on one in mainstream classroom

Medical Team

- Allergy
- Audiology
- Behavioral Health
- Cardiology
- Endocrinology
- Neurology
- Oncology
- Ophthalmology
- Otolaryngology
- Rheumatology

Otologic Recommendations

- Otologic management
  - Prescribed a trial of high dose prednisone and consult with his endocrinologist regarding cortisol levels
  - Recent MRI was consistent with normal inner ear anatomy and no changes in brain
  - Audiologic re-evaluation in 2 weeks
Audiologic Recommendations

- Repeat everything
- "Red Flags" that this is not a typical case
- Consistent responses but needed encouragement
- Lack of social engagement during testing
- Speech studies do not correlate with pure tone findings

Audiologic Results: 7/28/08

How Should I Proceed?

- Moved forward with amplification utilizing our department's hearing aid loaner bank
- Patient and family were very motivated
- Hesitant to have family purchase hearing aids until we have established stable hearing sensitivity
Fluctuation Patterns

- Decreased to severe hearing loss range in November 2008 following otitis media requiring antibiotics
- Following another round of oral prednisone hearing sensitivity improved to mild to moderate hearing loss range by end of December
- Autoimmune work-up due to recovery with steroid treatment
  - Negative results but prescribed methotrexate

Effects of Prednisone

Pros
- Improved pure tone thresholds
  - Improved attention at school
  - Able to participate in conversation at home
  - Mom reports he is overall “engaged in his environment”

Cons
- Significant changes in behaviors and personality
  - Food seeking habits
  - Difficult to control
  - Running away from home
  - Insomnia
  - Self harm
  - Hospitalization

Hearing Aid Evaluation

General Considerations
- Flexible fitting range
- Volume control
- Multiple programs
- Remote control option
- Hearing journal/tracking

Individual Considerations
- Phonak Naida SP
- Volume control & program button disabled
  - OCD behaviors
- Open lines of communication
August 2010

- Parental concern
  - Future fluctuations
- Physician uncertainty
  - 2nd opinion
- Time to take another look
  - What have we missed?

Audiologic Results:
8/19/10

Why ABR? Why Now?

- Physician request for objective testing
  - Prove that fluctuations were real
  - Rule out concerns of malingering / behavioral inconsistencies
  - Wanted to rule out AN/AD
  - Looking past inner ear into possibility of increased intracranial pressure given medical history
Testing Complications

- Request to compare same day audiogram to ABR
- Two attempts to complete an ABR
  - Natural sleep
  - Natural sleep with prescribed nightly sleeping pill
- Neither appointment was successful
  - Audiologic necessity vs. medical necessity

Pre-ABR Audiogram

ABR Testing

- ABR was performed under general anesthesia on November 19, 2010 in conjunction with a MRI and LP
- MRI revealed normal inner ear anatomy, no enlargement of the vestibular aqueduct, and stable brain findings
- LP revealed normal pressure levels ruling out increased ICP or pseudotumor
ABR Results

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<th>500 Hz</th>
<th>1000 Hz</th>
<th>2000 Hz</th>
<th>4000 Hz</th>
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<tr>
<td>Right ear</td>
<td>65</td>
<td>70</td>
<td>75</td>
<td>70</td>
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<tr>
<td>Left ear</td>
<td>75</td>
<td>60</td>
<td>75</td>
<td>70</td>
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500 Hz Air Conduction

1000 Hz Air Conduction
Additional ABR Results

- No response to unmasked bone conducted 2000 Hz and 4000 Hz tone burst stimuli at 40 dB nHL
- Absence of identifiable cochlear microphonic to air conducted click stimuli at 90 dB nHL
Comparison of Results

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<tbody>
<tr>
<td>Right ABR</td>
<td>65</td>
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<td>70</td>
<td>65</td>
</tr>
<tr>
<td>Left Audio</td>
<td>60</td>
<td>65</td>
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Where Are We At Today?

- Hearing continues to fluctuate between moderately severe to profound range
- Most recent audiograms on 1/20 and 2/21 showed SNHL in the severe range

Recent Audiograms

1/20/11

2/21/11
Where Are We At Today?

- Increased parental frustration
- Quality of life concerns
- Cochlear implant?
- Minimal improvement to steroid treatments
- Autoimmune testing negative, normal LP level, ruled out pseudotumor

How To Monitor These Patients?

- Regular appointments
- Search for pattern
- Utilize all tools in your diagnostic arsenal
- Flexible amplification
- Trust your professional judgment
- Consult with other professionals

Melissa L. Caine, AuD
The Children’s Hospital of Philadelphia
267-426-0264
caine@email.chop.edu
Audiologic Cross-Check & Other Tricks Up Your Sleeve

Violette Lavender, AuD

Objectives

- Identify audiometric "red flags" of central auditory pathologies.
- Cross check testing to aid in the differential diagnosis of central vs. peripheral auditory/vestibular pathologies.
- Recognize when to refer out for specialized testing in the differential diagnosis of auditory/vestibular pathologies.

Case

18 year old boy presents to our clinic for a CI consultation and vestibular evaluation
- History of Ototoxicity
History

5 YEARS AGO:
- Headaches, ataxia
- MRI: Brainstem glioma with focal lesions involving thalamus, cerebellum, and spinal cord
- Lesion biopsy, rendered him unable to walk
- Malignancy confirmed
- Inoperable, tx with chemo and radiation
- Last round of chemo 6 months ago

FOLLOWING TREATMENT:
- Tumors had shrunk considerably
- Physical therapy helped him walk again independently and with a walker for long distances
- VP shunt on the right side to manage hydrocephalus
- G tube for feeding due to swallowing abnormalities
History

...NOW
– Slow decline in balance, hearing, and vision

Rotary Chair

• Abnormally Low Gain- most often aligns with a bilateral vestibular weakness

Vestibular Evaluation

• Absent Caloric Responses to Ice Water
Vestibular Evaluation

- **Ocular-Motor:**
  - Smooth pursuit tracking: low gain & leftward direction weakness.
  - Random saccade testing: rightward direction undershoots (long latencies) & slow velocities in both directions.
  - Horizontal gaze testing: RB nystagmus looking right and LB nystagmus looking left

Gaze Testing

- Looking left = left beating nystagmus
- Looking Right = less intense right beating nystagmus
Smooth Pursuit Tracking

- Note the “saccadic” pursuit

Positional Testing

Differential?

- Bilateral vestibular weakness
- Upper brainstem pathology
- Cerebellar pathology
Audiologic Evaluation

- Absent ipsilateral and contralateral acoustic reflexes

Differential?

Auditory Processing Disorder
Auditory Neuropathy/ Dysynchrony
Neoplasm status change
Stroke
VP Shunt malfunction
Others?
ABR

- DPOAE's present bilaterally
- ABR:
  - **Right Ear**: Wave I was present at a high intensity level only. No other identifiable waveforms present. Waveform morphology was poor.
  - **Left Ear**: Wave I, III, and V were present at a high intensity level. Wave III and V were both delayed. Wave V diminished with the use of a faster click rate. Waveform morphology was poor.

**Differential?**

- Upper Brainstem Pathology, worse on the right side, cerebellar involvement based on vestibular findings