Co-treatment in Complex Cases: The Intersection of Audiology and Speech-Language Pathology

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Disclosures

NONE
Outline

• Complex case (medical and audiologic)
• Medical diagnosis
• Audiologic presentation
• Team approach to decision making and management
Cara’s Story
(used with her family’s permission)

• Cara was born full term without complications
• Passed newborn hearing screen with otoacoustic emissions (DPOAEs)
• Normal developmental milestones in first year of life
Background

At 15 months:
- Cara developed URI with fever;
- One day later, she woke up from a nap and had abrupt onset of vertical nystagmus.

At 16 months:
- Ophthalmic exam:
  - Electroretinogram consistent with outer retina pathology
  - Began showing unsteady gait and hand tremors
  - Treated for 2 months with intravenous corticosteroids, immunoglobulin since neurologists thought she had an autoimmune disease
  - Transient improvement in nystagmus, gait abnormality but each time symptoms recurred

By 17 months:
- Weakness of both upper extremities, increased gait unsteadiness, head tilt to the right side, excessive drooling
- Hospitalized for further evaluation
Medical Evaluations

• CT with contrast of chest, abdomen and pelvis
• MRI with and without contrast of the head and cervical spine
• 24 hour urine for catecholamines (hormones found in urine; used to dx rare tumors of adrenal glands, e.g. neuroblastoma)
• EEG
• Lumbar puncture was performed
• All tests were negative
Background

• By 19 months:
  • Child had shown no sustained response to steroid tx
  • Given rapidly progressing neurodegenerative symptoms, therapy with cyclophosphamide, a chemotherapy agent, was considered as next possible step.

• At same time:
  • Expedited genetic analysis was initiated given possibility that a genetic diagnosis might change the treatment plan. This was done as part of a research collaboration.

Petrovski et al. 2015
Whole-Exome Sequence Results

Genomic Analysis:

• Revealed progressive neurodegenerative disease called Brown-Vialetto-Van Laere Syndrome 2 (BVVLS2) that is a consequence of severe riboflavin deficiency; aka riboflavin transporter disorder (RTD)
• Untreated BVVL is a rare and often fatal disorder
• Previously, no effective treatment
  • But in past few years researchers discovered that high doses of vitamin B in some patients have had dramatic effects on halting progression of disease, especially when started early in disease course
• All patients diagnosed with the disorder have auditory neuropathy type hearing loss.

Cara’s Treatment:

• At 20 months of age, Cara began receiving high dose oral riboflavin three times/day.
  • Parents reported improvement in her function within a week of first dose; drooling less, less head tilt
Meet Cara
Audiologic test results

• Family requests Audiology consult at UNC since they had been told that most patients with BVVL have auditory neuropathy and hearing status remains unclear

• Due to child’s compromised respiratory function, sedated ABR had not been possible earlier and incomplete information had been obtained using behavioral audiometry.

• ENT exam showed presence of middle ear fluid so tubes and ABR were recommended.

• Results of ABR showed absent neural responses at high intensity levels with present cochlear microphonics and present OAEs consistent with auditory neuropathy.
• SRT:
  • Right: CNT
  • Left: 25dBHL

• Recorded PBKs:
  • Right: CNT
  • Left: 48%

• Tympanometry
  • Consistent with patent tubes

Initial behavioral audiometry at age 4 1/2
Hearing Aid/Remote Mic Fitting
Age: 4 ½ years

• Given extremely poor word recognition in right ear, decision made to proceed with hearing aid fitting with integrated FM (Roger) receiver for left ear only
• Parents purchased HA and receiver, school provided transmitter for school and home use
• Initial impressions by parents and teachers regarding HA and remote microphone system were positive
Follow up

• Ultimately, despite use of HA and remote microphone (Roger) system in left ear, child continued to require frequent repetitions.

• Parents have communicated with other families with same dx whose children have received CI and wonder if she is candidate for CI in right ear.

• At 4 yrs 9 months, child referred for CI consult
CI Consult

Speech perception scores (PBK,MLV):

- Left (aided): 52% words, 77% phonemes
- Right (unaided): 24% words, 53% phonemes

- After discussion and extensive counseling with family and CI team members, decision made to proceed with right CI
- Child received right CI at age 5 yrs 4 months
Decision to proceed with CI difficult:

Child Factors:
• Child’s attention span for testing was limited with inconsistent results
• Significant amount of residual hearing in both ears by pure tone testing
• Effects of B2 vitamin therapy on auditory system unknown

Factors related to disorder:
• Rare disorder with few published reports re hearing outcomes
• Only a few case study reports in literature with mixed outcomes for CI
• Questions re effect of presumed post-synaptic site of lesion

Rance and Starr 2015:
Unaided pure tone thresholds and CI detection thresholds right ear (7 months post-op)

- PBK, MLV, Right (CI):
  - 78% words; 93% phonemes
Family Decision re Left Ear CI

• Following several months of CI use in right ear, phone call received from family saying child now rejecting left HA; return appointment scheduled for follow up and further testing.
• Ultimately family decided to proceed with second CI
• As of this presentation, child has only been activated about 2 months so limited information regarding CI outcome available but family and therapists initial reports are positive.
What Does It Mean to Collaborate?
Co-Treating at UNC

• Collaboration

• Family Support

• Whole Team Communication

• Collecting Data and Coaching towards Goals
Co-Treat Follow Up for Cara

Initial Visit & Prior to CI

Post Cochlear Implant
Therapy & CI Ear Isolation

Evans & Hancock, 2019
Speech, Language & Functional Auditory Skills
Speech & Language Results

Pre CI (aided left ear only)
CA: 5.0
*Scores interpreted with caution as multiple repetitions needed

Post CI (unilateral CI, no HA)
CA: 6.3 Years  CI Age: 1 year
Functional Listening Index (FLI-P)

First CI 63 Months

Second CI 72 Months

Shepherd Center, 2013
Subjective Reporting from Cara’s Parents

- 5 Week’s after Activation
- 3 Month’s post 1st CI
- 5 Month’s post 1st CI
- 2 Month’s post 2nd CI
Key Points

• Collaboration is the key for all children considering cochlear implantation but especially in complex cases like this one

• Multiple specialists involved in child’s care with multiple visits and frequent conversations needed:
  • Ophthalmology, neurology, genetics, ENT, Audiology, speech pathology, educational professionals.

• In this case, whole exome sequencing resulted in a transformative change in child’s medical management.

• Brown Vialetto Van Laere or other progressive neurologic diseases should be considered in differential diagnosis for children who present with ANSD
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Thank you

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www.ci2020orlando.org
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