

Autism Spectrum Disorders in Children who are Deaf/Hard of Hearing



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Learning Objectives

- Describe aspects of atypical development in children with the dual diagnosis
- Identify communication strategies which can help with children with a dual diagnosis
- Gain an understanding of family perspectives regarding the dual diagnosis of hearing loss and autism spectrum disorders

Impact of UNHS/EHDI on language outcomes

- Universal Newborn Hearing Screening has allowed the earlier identification of children with hearing loss
- This subsequently enhances age of intervention with data to support language outcomes in the average range for early identified children
- With this improved language trajectory, we should be striving for earlier identification of children with co-existing autism spectrum disorders, however diagnosis can be challenging due to a lack of validated tools

Why is it important? ASD and HL

When there are **two** reasons for communication problems and those problems span varying expertise (teachers of the deaf, educators with expertise in autism)....

it can be very challenging to implement effective strategies to develop communication and social interaction

How common is it?

Epidemiology of HL

- 2-3 per 1000 with congenital moderate to profound hearing loss (MMWR 2006)
- 6 per 1000 with mild to profound hearing loss by the age of 6

Epidemiology of HL and ASD

- Estimate of 1-6% of children who are deaf/hoh also have an ASD
 - Rosenhall: How many children with ASD have a hearing loss: mild to moderate 7.9%, profound 3.5%
 - Gallaudet Research Institute: How many deaf students have ASD: 1%
 - Jure et al: How many deaf students have ASD: 4%
 - Syzmanski et al 3.4%

Challenges

- Age of diagnosis of an ASD tends to occur at older ages (>3 years – 15 years)
- Comorbidity of ASD and hearing loss significantly complicates language development
- Lack of validated evaluation tools and expertise, however it is inappropriate to make a diagnosis of ASD without relying on standardized measures (can cause over-diagnosis and under-diagnosis)
- Management of hearing loss and communication needs complex

Core Deficits

Atypical Communication

- Not just delayed language, but atypical use, such as echolalia, scripted language, poor use of gestures, difficulty sustaining conversation, a lack of social and imitative play

Social interaction

- Difficulties with eye contact, joint attention, facial expression, understanding of gestures and non-verbal communication, lack of interaction with peers, lack of social reciprocity (showing, shared enjoyment)

Restrictive, Repetitive Interests

- Intense interests or preoccupation, tend to be unusual interests, adherence to routine or ritual

Deaf/HOH Child's Processing

- A deaf child must:
 - divide attention,
 - retain information in memory,
 - integrate the information from the activities AND the language input
- Added burden if a child is relying on lip-reading to any extent

When might you suspect ASD in a child who is deaf/hoh?

- Problems in pre-linguistic communication

- Joint Attention
- Eye Contact
- Requesting
- Showing
- Turn-taking
- Choice making

May not understand

Lack of clarification strategies

Not aware of misunderstanding

Difficulty integrating information

Question-asking skills limited

Reduced range of language functions

Gaps in social understanding

- Influences reasoning

Pre-Linguistic Communication Red Flags for Autism

Barbaro, J. and Dissanayake, C. **Prospective Identification of Autism Spectrum Disorders in Infancy and Toddlerhood Using Developmental Surveillance: The Social Attention and Communication Study.** Journal of Developmental & Behavioral Pediatrics. 31(5):376-385, June 2010.

Behavior	8 months	12 months	18 months	24 months
Eye Contact	X	X	X	
Turning to Name Call	X	X		
Imitation				X
Pointing		X	X	X
Gestures-Waving		X	X	X
Pretend Play			X	X
Showing Behaviors				X
Fail Criteria	Fail 2/2	Fail 3/4	Fail 3/4	Fail 3/5

Retrospective Chart Review

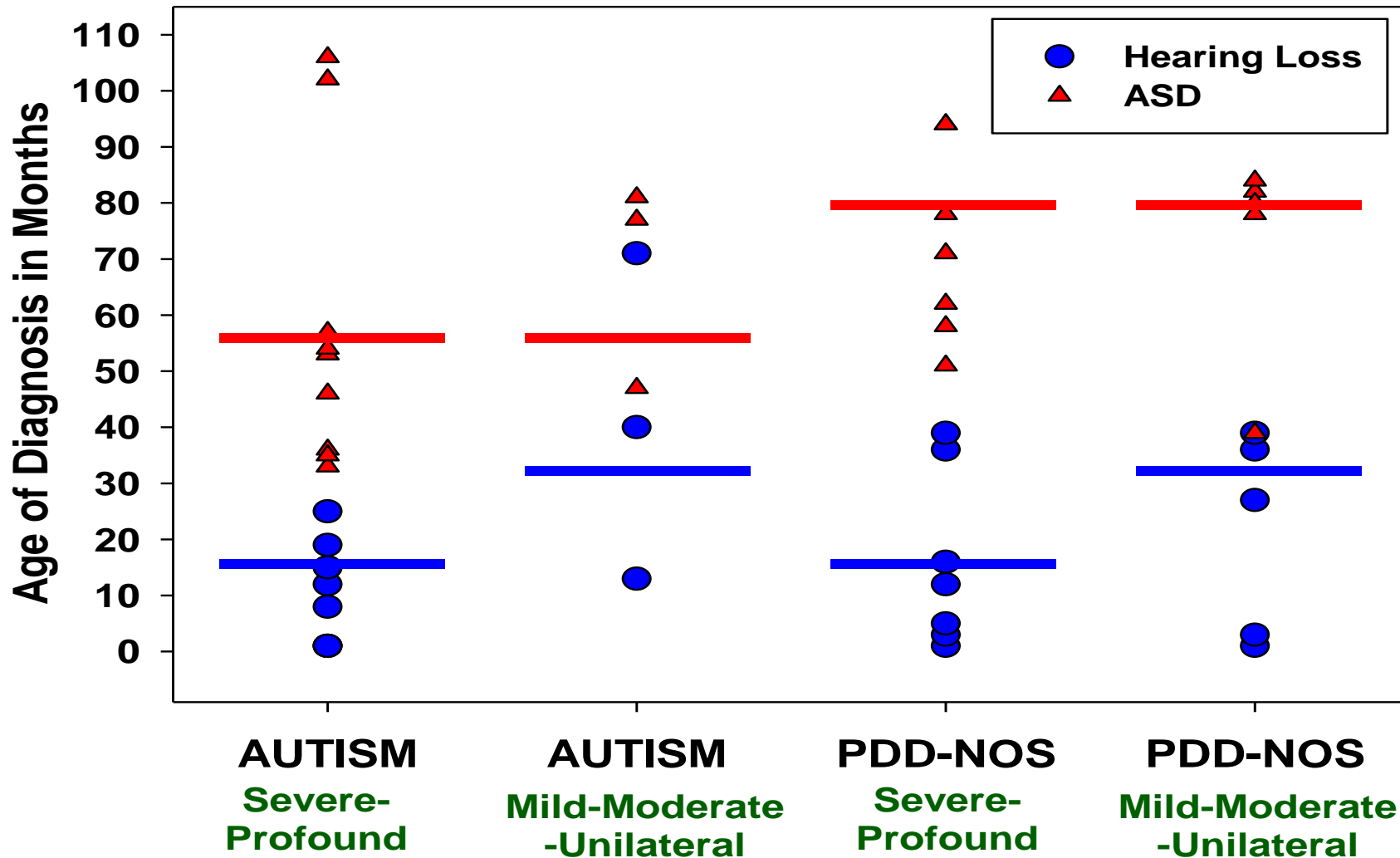
- Study Objective: To describe a population of children with the dual diagnosis of permanent hearing loss and ASD in the context of the degree of hearing loss as well as the ASD diagnosis
- 24 children of varying degrees of HL and ASD

Meinzen-Derr, J et al “Autism Spectrum Disorders in Children who are Deaf or Hard of Hearing” International Journal of Pediatric Otorhinolaryngology 2014 Jan;78(1):112-8.

Age of Diagnosis for ASD and Hearing Loss

Median 53.5 months

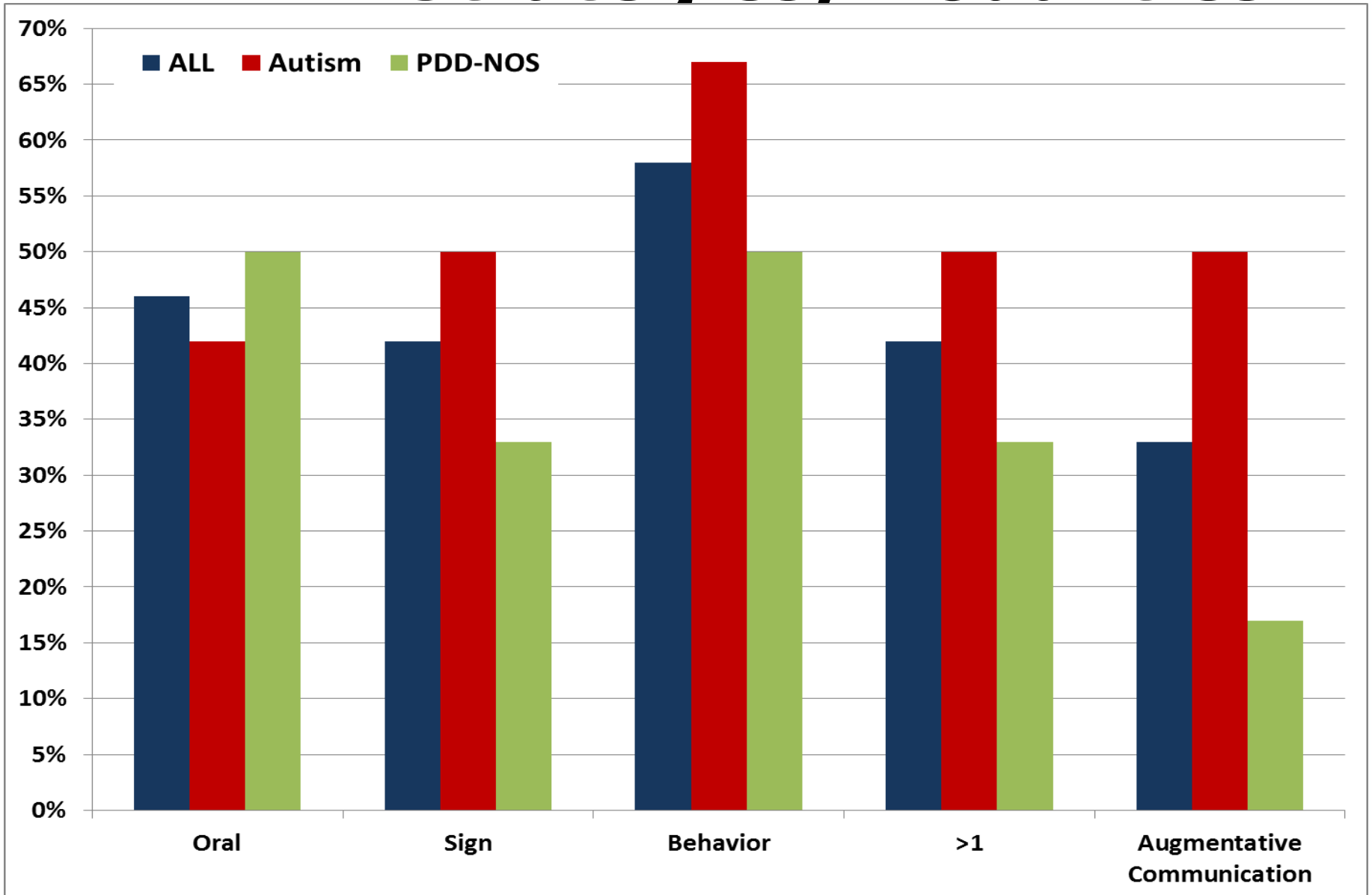
Median 78 months



Earlier Ages of ASD Diagnosis

- More severe hearing loss (earlier ages)
- More severe ASD (Autism vs. PDD-NOS)
- Not associated with:
 - Etiology or amplification
- Not correlated with:
 - age of amplification ($p=0.4$), nonverbal cognitive abilities ($p=0.7$), language abilities ($p>0.3$)
- Did not review age of suspicion!

Communication Strategies/Modalities



Drivers of communication abilities

- The nonverbal cognitive quotient was significantly correlated with both receptive ($\rho=0.76$, $p=0.003$) and expressive ($\rho=0.83$, $p=0.003$) language levels
- Children with ASD diagnoses of PDD-NOS and Asperger's had higher language scores relative to their cognitive abilities
- In addition, children with mild, moderate, or unilateral hearing loss had higher language scores relative to their cognitive abilities compared to those with severe to profound hearing loss
- The ages of identification of hearing loss and diagnosis of ASD were not significantly associated with either receptive or expressive language levels ($p>0.3$ for all correlations)

“Success” defined

a : degree or measure of succeeding

b : favorable or desired outcome;

also : the attainment of wealth, favor, or eminence

- Merriam-Webster Online

How do you want to define “success”?
How do the parents want to define “success”?
Do they match up?

Issues to Consider

1. Some disabilities are not easily identifiable at the time of consideration for candidacy
2. In more complex cases, hearing loss may not be the first priority
3. Even in children without additional disabilities, outcomes depend on other factors such as chosen mode of communication

“Despite the best efforts of many professionals, it is often difficult to diagnose learning disabilities, reduced cognitive function, and soft neurologic deficits in very young children...”

-Walzman, 2000

Special Populations

- 40-50% of children with hearing loss will have an additional disability (Wiley, et al., 2004)
- Given the benefits of early intervention, there will be children who receive hearing intervention prior to the identification of additional disabilities
- Disabilities such as autism or apraxia may not be identified until a child is at least 2-4 years of age
- A language, learning, or cognitive disorder will still be present after a child receives hearing device. It is important that everyone understands that the device is not going to resolve all issues
- Children with additional handicaps *need* an experienced multi-disciplinary team to assist in determining appropriate expectations
- Realistic and appropriate expectations must be discussed and understood by the family and professionals involved.

Audiologist Challenges

- Uncomfortable with very involved children
- May not realize child's potential
- Providing education to actual caregivers



Diagnosis Challenges

Delay in Diagnosis

- Other medical issues may take priority
- Parents may already be overwhelmed
- Hearing loss is not a visible disorder
- Child may be difficult to test
 - Developmental ability of child to perform test
 - Concern for sedation
- Middle ear dysfunction

Diagnosis Challenges

Delaying Other Diagnosis

- May be so focused on hearing loss that other diagnosis may be delayed
 - Autism spectrum
 - Speech/Language impairment
- Need to look beyond auditory behaviors and know when to refer for other services
 - Developmental Pediatrics
 - Occupational Therapy
 - Speech Therapy

Amplification Challenges

- Late intervention
- Inconsistent test results
- Medical issues may take priority
- Yet another piece of equipment
- Sensory integration issues
- Unwanted environmental sounds
- How to determine benefit?



Evaluation:

Inconsistent Test Results

- Child may not be interested in traditional test stimuli (speech/warble tones)
- Lack of ear-specific behavioral thresholds
 - Could easily miss unilateral hearing loss
- Neurologic insults and ANSD can result in abnormal ABR results
 - Behavioral testing becomes very important
 - Parents can be key to knowing what is a response in their child and what sounds may interest their child

Evaluation:

Other considerations

- Evaluate for communication ability, not just hearing sensitivity
- Does this child make use of the information he receives from his intact sensory modalities?
 - Environmental involvement – vision and touch
 - Does the child accept or reject this input?
 - How might this relate to tolerance of device wearing or the stimulation it provides?

Beyond audiograms and speech perception measures...

- **Some thoughts and questions to ponder:**
 - Is there additional information that should be considered for closer analysis?
 - Are we already getting the information without evaluating it's value?

Management Tips and Tricks

Understanding the Child

- Really take time to get to know the child...
- Follow the child around the clinic for an hour or so
- Let the child show you what he/she is comfortable with
- Involve an common or established party (therapist, child life specialist, teacher, etc.)
- Consider the most appropriate time for the child's schedule, and not the clinic's schedule
- Pre-Medication? (ADHD, anxiety, others?)

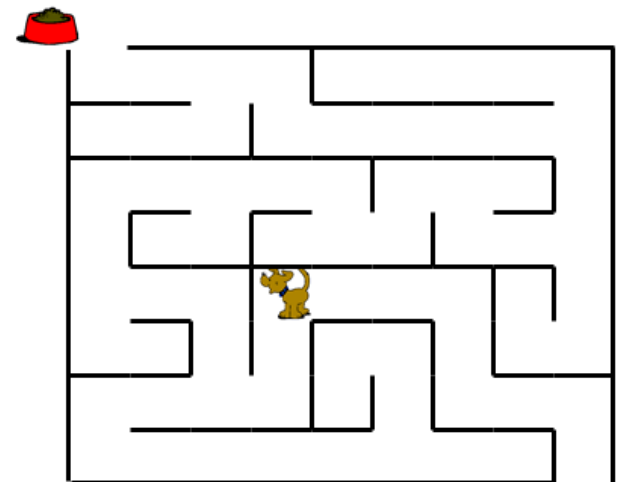
Management Tips and Tricks

New Games

Get the parents/caregivers to help out

- Ask the parent what entertains their child
- Teach the parents how to play a listen-n-drop game at home that you could also play in the office
- Make up new "games" →

Flexibility is key!



Computerized Conditioned Play
-courtesy of Boys Town

Management Tips and Tricks

Environment

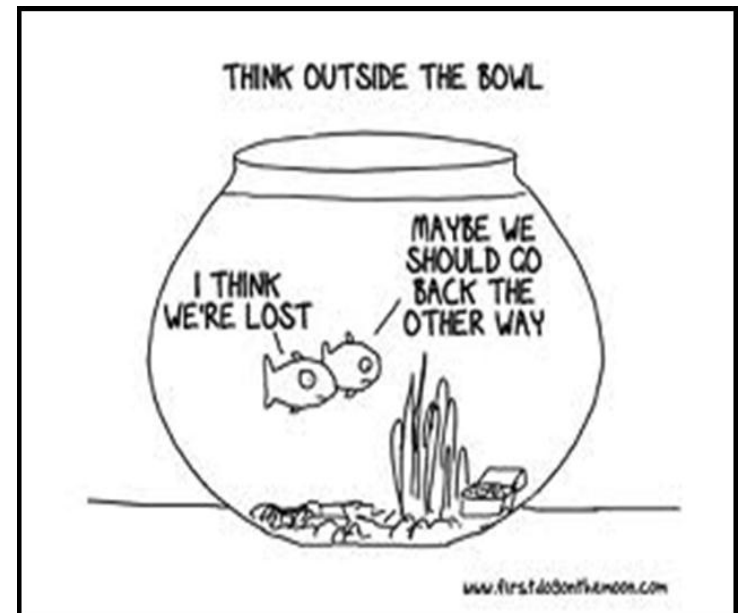
- Have a variety of toys for possible tactile issues
- What might be a distractor in the environment?
 - Pictures, words, numbers, lighting?
- Consider the chair you are using:
 - a chair that spins/swivels for the child who needs that extra stimulation to stay on task
 - a chair with a foot-rest for the child that needs to feel “grounded”

Management Tips and Tricks

“Environmental Responsiveness”

Go to where the patient is most comfortable

- The child may give you more feedback and/or be more willing to play listening games in a familiar environment such as:
 - the place he has weekly therapy
 - school environment
 - elevator? - in jest, but think outside the box!



Sensory Integration Issues

- Consider retention accessories as well
- Start with small intervals of use
 - May tolerate better once benefit is noticed
- Parental perseverance
- Work closely with occupational therapy

Management Tips and Tricks

The Device

- With a cochlear implant, perhaps you can only get the coil on, so the processor has to hang off of the ear for a while- and that's ok!
- Try putting the device on without batteries in (no auditory stimulation) first, only to work up to consistent wearing followed by progressive programming
- Take baby steps and build upon the encouragement of small success

Management Tips and Tricks

Active Involvement

Involve the child more than you typically would:

- Show him the programming screen
- Show him that he can hear a sound when he sees something change on the screen
- Of course, stop short of handing him the mouse and keyboard!
- Sometimes, the child just needs to make a connection between what they are hearing and what they see

Management Tips and Tricks

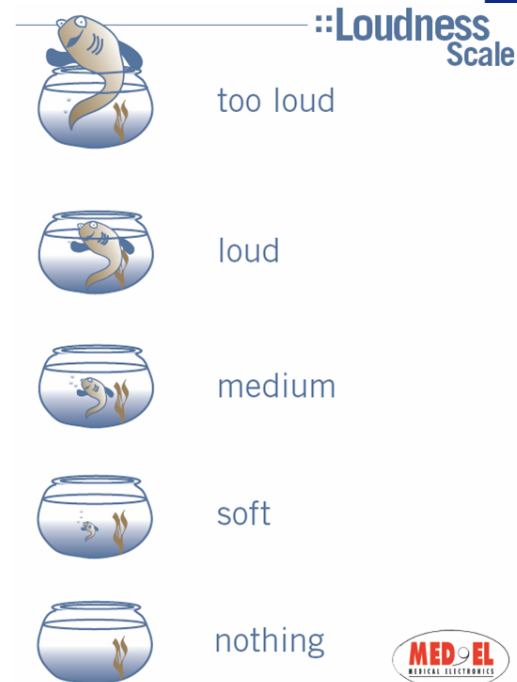
Verification during programming

Objective Measures:

- Hearing aids: simulated and/or real ear measures
- Bone conduction devices: bone simulator measures
- Cochlear Implants: impedances, telemetry, stapedial reflexes

Subjectively:

- Sweep across the upper levels and observe reaction
- Make various sounds and carefully observe reactions with use of the new program
- Try loudness scaling- you may be surprised!
- Adjust accordingly



Management Tips and Tricks

Determining Benefit

- Traditional aided testing may not be possible
 - Use familiar toy, voice, CD or video
- Functional outcome measures may help
 - ELF, LIFE, CHILD, IT-MAIS, MAIS, etc
- Ling Sound Test
- Keen observation of the child during programming
- The audiologists' past experience with other patients
- Parent/therapist/teacher reports



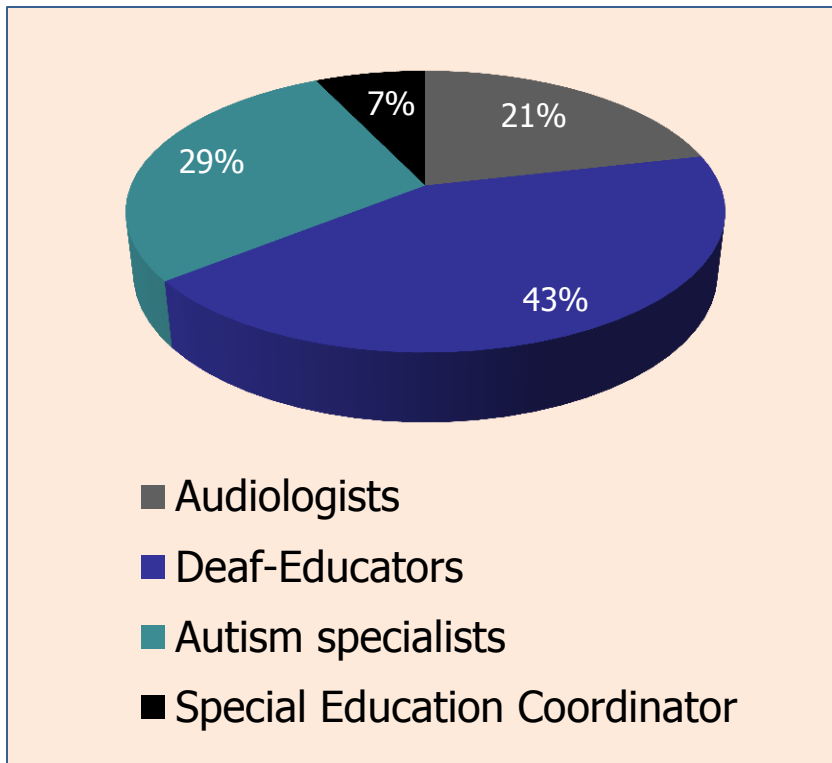
After the Fact: Outcomes

“Although the development of auditory skills was not as rapid or favorable for these subjects...the children in this study did obtain benefit. In addition to improved auditory skills, communication skills, social interactions, and general “connectedness” to the environment increased.”

-Waltzman, (2000)

Focus Group Professionals

Objective: To understand the involvement of the professionals who work with children with the dual diagnosis and their families.



Invited participants based on their expertise in either the field of deaf education/hearing loss, the field of autism spectrum, and those who have worked with children with known dual diagnosis.

15 participants

Topics of Interest

- Useful tools
- Educational options
- Choosing an educational setting
- Working on the child's team
- Ideal program

Discussion Themes

- Educational Settings
- Medical & Educational Teamwork
- Family Education
- Role of ASL Interpreters

Educational Settings

- Efficacy of education setting is child dependent
 - Focus on behavior/ability that is most interfering at the time
 - Requires many different resources and providers
- Peer communication is essential
 - Deaf Education programs often better facilitate communication in social settings
- Visual supports have been most effective
 - Tactile/hands-on education is motivating

Medical and Educational Teamwork

- Participants unanimously agreed that the most important aspect of better serving children with PHL and ASD is collaboration between the medical, therapeutic, and educational settings

Medical & Educational Teamwork

- Cross-discipline training
 - Understanding the impact of the second diagnosis
 - United front for recommendations to the family
 - Children may perform differently in different settings, limiting our understanding of the child's needs
- Cross-discipline communication
 - Best method?
 - May not be the same for all types of families
 - Necessary information?
 - Not a billable service

Family Education

- Stronger understanding of communication basics
 - Impact both PHL and ASD have on the communication development of their child
 - Strengths and challenges within each education setting available
- Improved education may help parents be better consumers of health information
 - Improved trust in the child's health, therapy, and education team members
 - May encourage them to act as an equal member of team

Focus Group: Parents

Objective: To gain an understanding of the experience of families with children who have PHL with a co-existing ASD.

- Recruited families from around the region based on children who had received a dual diagnosis through CCHMC.
- Of the 16 families invited, five RSVP'd, but only three families attended (2 of 4 parents were fathers)

Wiley, S, Gustafson, S, Rozniak, J. "Needs of Parents of Children who are Deaf/Hard of Hearing with Autism Spectrum Disorder" The Journal of Deaf Studies and Deaf Education 2014 Jan;19(1):40-9.

Children of Participants

Children varied in degree of hearing loss and in ranges of severity of ASD

Characteristic	Child 1	Child 2	Child 3
Type and Degree of HL	Unilateral mild to moderate	Profound bilateral	Profound bilateral
Age of HL identification	3 years	2 months by UNHS	Birth due to Syndrome
Type of Amplification	Hearing Aid	Bilateral CI	Unilateral CI, no longer using
Autism spectrum label	PDD NOS	Autism	PDD NOS
Age of Identification of ASD	6 years	4 years	6 years

Children of Participants

Characteristic	Child 1	Child 2	Child 3
Reminder	Unilateral, PDD-NOS	Profound bilateral, Autism	Profound bilateral, PDD-NOS
Current Educational Placement	Mainstream setting with IEP supports	Social Communication Classroom	Autism Classroom
Early Educational Placements	Special Needs Preschool	Home-based EI School for the Deaf with manual approach in apraxia program	Home-based EI Classroom for children with motor disabilities Special Needs Preschool Developmental Disabilities Classroom
Communication Mode	Aural/Oral	Total Communication: writing, signing, spoken language	Signing
Autism specific strategies used	Visual Schedules Social Stories	Visual Schedules PECS	Visual Schedules, PECS, Dynavox

Topics of Interest

- Experiences during diagnosis
- Impact of dual-diagnosis on family
- Useful resources
- Wish-list

Discussion Themes

- Diagnostic Experiences
- Family Impact
- Functional Skill Development
- Academic considerations

Functional Skill Development

- Functional skills are a priority
 - Struggle to balance development of social skills and academic performance
 - reported more interest in the development of their child's social and functional skills

*“At some point, I’m going to look at him and say, it doesn’t matter to me you know your multiplication table, I can’t take you anywhere.
I can’t take you to the grocery store.”*

- Access to social groups is a challenge
 - Accessible social groups based on geographic location and scheduling convenience are difficult to find within already busy schedules

Academic Considerations

- Parents were happy with services that were provided in school
 - Concerned about potential of cuts in funding
- Limitations of Extended School Year
 - Need continuation of the academic school year's structure on a year-round basis, particularly related to their child's difficulties with changing schedules and reliance on structured teaching
- Requested better training of professionals
 - Ensure that professionals working with their children are proficient in both PHL and ASD

Useful resources

- All families noted that schools were primary resources for information and support.
 - Important to have strong communication between school and home
 - Allows consistent follow-through for supporting their child's development at home

“It all depends on your teacher. We used to get materials to bring home that we could work on with her on vacations and breaks. Now we aren't getting any of that.”

Useful Resources

- Visual schedules
- Internet was a consistent resource used by all families to:
 - Learn more about strategies to help their children
 - Network with other families
 - Connect with professionals to provide guidance with problems

“The internet has been a godsend, I couldn’t do anything without it.”

Wish-list

- Ability to observe their child's out-patient clinical therapy.
 - E.g. video monitors
 - Thought to be helpful in allowing them to observe interventions to try at home with their child
- Someone that they can call/email with questions.
 - Contact should have a knowledge base of both PHL and ASD

What we've learned

- Families want more information
- Both professionals and families see better education of the medical and educational professionals as a priority
- Communication is KEY!

Closing Thoughts

- Collaboration
 - Professionals with experience in ASD, specific sub-population
 - OT, Behavior specialists
- Creativity
 - Think from the perspective of the child
 - Adapt known strategies to meet all of a child's needs
- Peer exposure
 - Define peer group, may need a variety of peer groups for different skills

Helpful Readings

- Odyssey: New Directions in Deaf Education: Autism Issue
- Spring/Summer 2008; Volume 9: Issue 1
- <http://www.gallaudet.edu/documents/clerc/odyssey-2008-v9i1.pdf>

Literature Review: General Articles

- Mandell et al Factors associated with age of diagnosis among children with autism spectrum disorders. *Pediatrics* 2005;116:1480-1486.
- Jure, R, Rapin, I and Tuchman, R Hearing-impaired autistic children. *Dev Med Child Neurol* 1991 33(12):1062-72.
- Rosenhall et al Autism and Hearing Loss. *J Autism Dev Disord* 1999 29(5):349-57.
- Worley, J, Matson, J, Kozlowski, A. "The effects of hearing impairment on symptoms of autism in toddlers" *Developmental Neurorehabilitation*, 2011 (14) 171–176.
- Szymanski, C, Brice, P, Lam, K, Gotto, S. Deaf Children with Autism Spectrum Disorders *J Autism Dev Disord* 2012 e-pub ahead of print Jan

Literature Review

- Gense and Gense: Autism and Deafness Workshop OCALLI 2002, 2005, 2007
- Guarinello et al Deafness and Attention in Deaf Children. American Annals of the Deaf 2007: 151:5:499-507.
- Peterson, C, Wellman, H, Liu, D. "Steps in Theory-of-Mind Development for Children With Deafness or Autism" Child Development 2005 (76) 502-517.

Literature Review: Assessment

- Johnson, K, DesJardin, J, Barker, D, Quittner, A, Winter, M. "Assessing Joint Attention and Symbolic Play in Children With Cochlear Implants and Multiple Disabilities: Two Case Studies" *Otology and Neurotology* 2008 (29) 246-250.
- Roper, et. al Co-occurrence of autism and deafness: diagnostic considerations. *Autism* 2003 7(3): 245-253.
- Hoevenaars-van den Boom, MAA, Antonissen, ACFM, Knoors, H, Vervloed, MPJ "Differentiating characteristics of deafblindness and autism in people with congenital deafblindness and profound intellectual disability" *Journal of Intellectual Disabilities Research* 2009 (53) 548-558.

Literature Review: Intervention Strategies

- Garcia, R and Turk J "The Applicability of Webster-Stratton Parenting Programmes to Deaf Children with Emotional and Behavioural Problems, and Autism, and Their Families: Annotation and Case Report of a Child with Autistic Spectrum Disorder" Clinical Child Psychology and Psychiatry 2007 (12) 125-136.
- Malandraki, G, Okalidou, A "The Application of PECS in a Deaf Child With Autism : A Case Study" Focus Autism Other Dev Disabl 2007 22: 23.

Literature Review: Outcomes Studies

- Donaldson, et. al. Measuring progress in children with autism spectrum disorder who have cochlear implants. Archives of Otolaryngology Head and Neck Surgery 2004;130:666-671.

Literature Review

- Gal, E, Dyck, M, Passmore, A. "Relationships Between Stereotyped Movements and Sensory Processing Disorders in Children With and Without Developmental or Sensory Disorders" *American Journal of Occupational Therapy* 2010 (64) 453-461.
- Maljaars, JPW, Noens, ILJ, Scholte, EM, Verpoorten, RAW, vanBerckelaer-Onnes, IA. "Visual local and global processing in low-functioning deaf individuals with and without autism spectrum disorder" *Journal of Intellectual Disability Research* 2011 (55) 95-105.

Literature Review: Family Perspectives

- Beals, K "Early Intervention in Deafness and Autism: One Family's Experiences, Reflections, Recommendations Infants and Young Children 2004 (17) 284-290.
- Myck-Wayne, J, Robington, S, Henson, E. Serving and Supporting Young Children with a Dual Diagnosis of Hearing Loss and Autism: The Stories of 4 Families. American Annals of the Deaf 2011 156:4, 379-390.