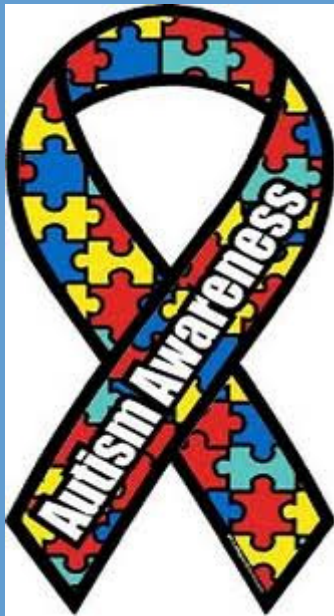


Autism Spectrum in Children who are Deaf/Hard of Hearing: Diagnostic and Intervention Conundrums



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Disclosures

- Neither we nor any member of my immediate family have a financial relationship or interest (currently or within the past 12 months) with any entity producing, marketing, re-selling, or distributing health care goods or services consumed by, or used on, patients.

Dr. Yoshinago-Itano is on the LENA Scientific Advisory Board but has no financial interest in the LENA Foundation

- We do not intend to discuss an unapproved/investigative use of a commercial product/device.

Learning Objectives

- Understand the higher rates of autism spectrum disorder in children who are Deaf/HH as compared to the general population
- Describe aspects of atypical development in children with the dual diagnosis
- Identify communication strategies which can help with children with a dual diagnosis

Why it is important

- At least 4% of children who are Deaf/hard of hearing (Deaf/HH) have an autism spectrum disorder which can further complicate communication development
- Although this rate is much higher than in the general population, the diagnostic process and availability of interventions to address communication challenges are severely lacking
- This disparity in access to appropriate diagnosis and intervention services greatly impacts possible long term improvement or outcomes in this group of children

Epidemiology

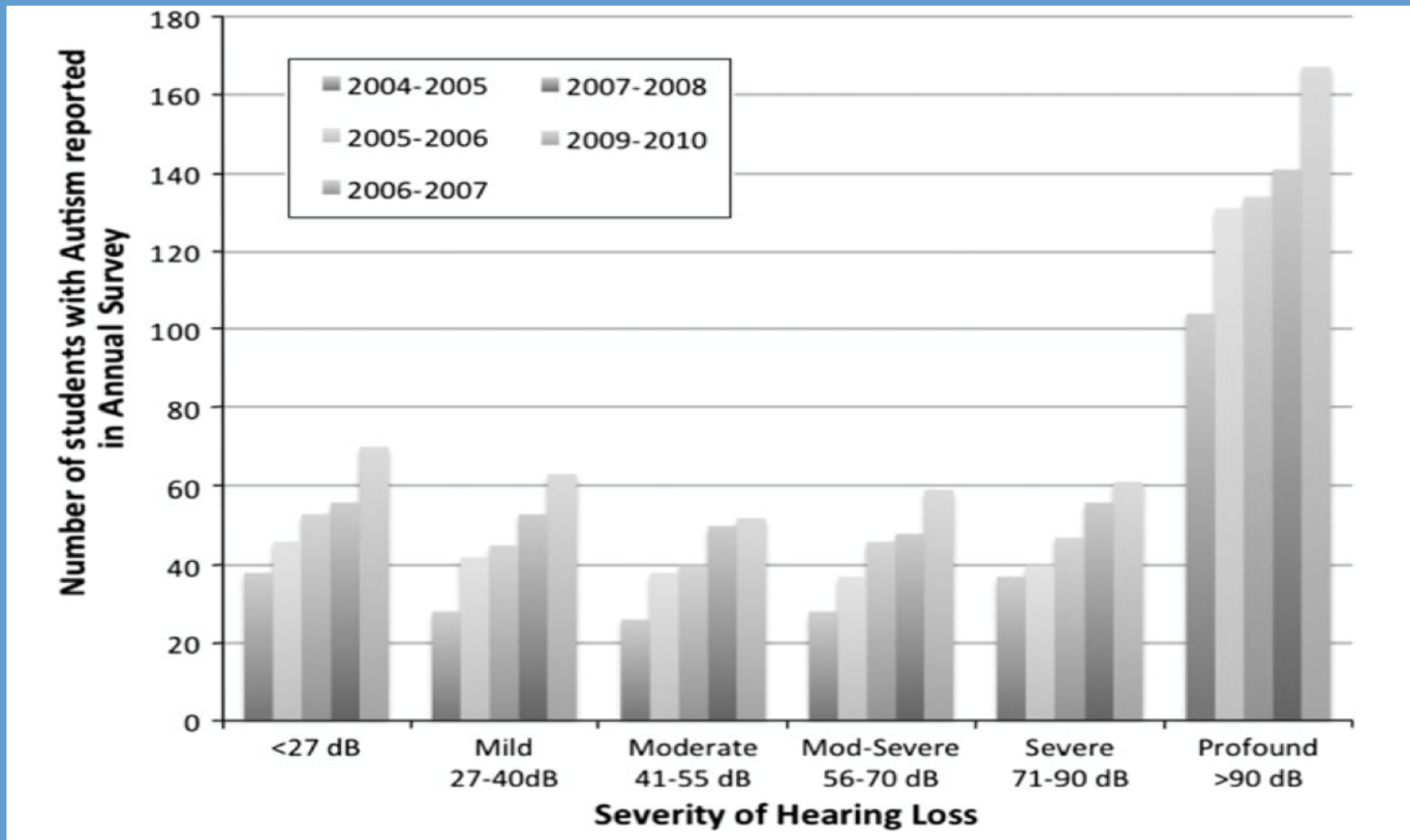
- Rates of ASDs continue to grow, even for children who are deaf

	CDC believed Prevalence Rates	Annual Survey believed Prevalence Rates
2004-2005	1:125	1:111
2005-2006	1:110	1:94
2006-2007	-	1:53
2007-2008	1:88	1:81
2009-2010	1:68	1:59

Understanding the dual diagnosis

- Previous research
 - Autism and severity of hearing loss are not related, but that may not be true
 - Congenital hearing loss should always be ruled out before an ASD diagnosis
 - Children who are Deaf/HH receive the diagnosis later
 - Autism and hearing loss are not related, but that also may not be true
 - Consistent rates that are almost always higher than children without hearing loss- current research

Prevalence of Autism based on Severity of Hearing Loss



Data is provided here from the Annual Survey of Deaf and Hard of Hearing Children and Youth Conducted by the Gallaudet Research Institute
Published in Szymanski, Brice, Lam and Hotto, 2012

The diagnosis: A dilemma?

- Dilemma of under-diagnosis
 - Is it the right one?
 - Lack of diagnosis = lack of early intervention = lack of possible gains later in life
 - Lack of appropriate services
- Dilemma of over-diagnosis
 - Is it the right one?
 - Supports and intervention that may not be appropriate may not help symptoms, nor prepare families or provide appropriate resources
 - Schools and professionals may not accept the child

Diagnostic Challenges

- Lack of standardized assessment tools for Deaf/HH
- Providers trained in deafness may not be trained in ASD and vice versa
- Providers trained in ASD may not be trained in understanding the complex needs of children who are Deaf/HH
 - Communication, development, behaviors etc.
- Children will act differently in different settings (multiple sources of information helpful)
- Assessment Challenges via interpreters
 - May not know/recognize or convey atypical language features if noted
 - Potential to disrupt rapport necessary for assessing social reciprocity
 - The role of an interpreter may be unknown

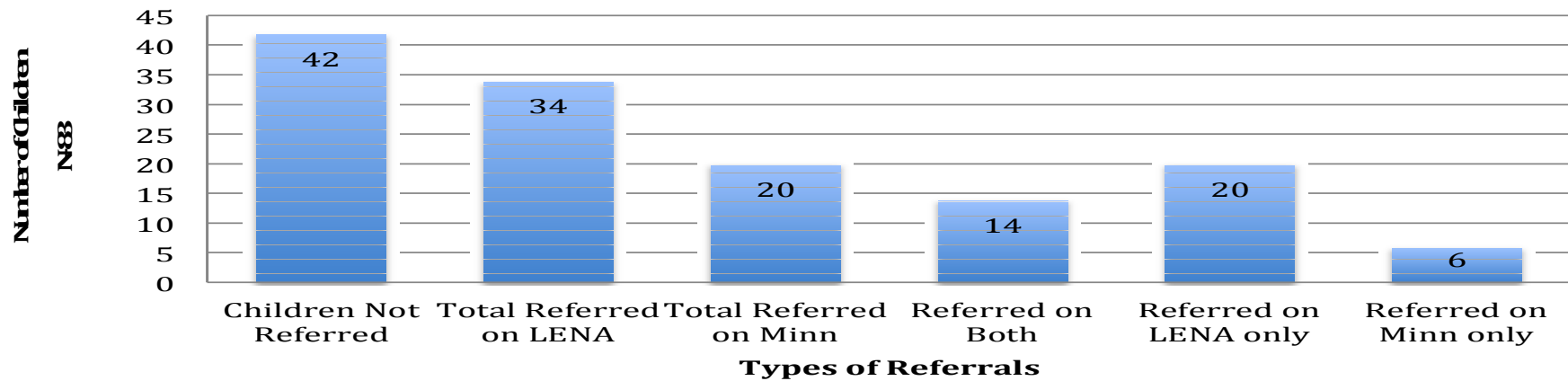
Screening

- No ASD screening tools have been validated for children who are Deaf/HH
- The Baby and Infant Screen for Children with aUtism Traits (BISCUIT) was administered to children with conductive hearing loss (need for PE Tubes) and children in this category did not overlap with the results from the children with ASD (suggesting that children who are at risk for HL do not have autistic traits)

Screening

- Colorado has applied the LENA autism screening (LLAS) among 83 children birth to 72 months who are Deaf/HH of varying degrees of hearing loss
- The LENA ASD screening algorithm was applied to the data outputs to categorize likelihood of an ASD
- Minnesota CDI results (specifically social development domain) were also evaluated among children flagged as at risk on the LENA
- Children at risk require further evaluation

Referral Rates for Criteria 3



Results

- “Criteria 3” is the most robust measure resulting in the most accurate need for referral.
- Using a double screen (LENA and CDI) the refer rate for the LLAS and M-CDI is 16.87%
 - Those that referred on LLAS but not the M-CDI was 24.10%
 - Those that referred on the MINN-CDI Social but not the LLAS were 7.23%
- Therefore, using a double screen relying on “criteria 3” is the most appropriate for determining who warrants referral for further evaluation as the other two criteria have a high false positive rate (indicate concern when no problems exist)
- The sensitivity for referral is robust for all types of hearing loss, except for bilateral severe/profound hearing loss

Results: Other Findings

- Among 20 children in the study flagged on the LLAS alone (not the social subscale on the MINN-CDI Social) did not have suspicions of ASD by their CHIP provider, suggesting further diagnostic evaluation may not be needed.
- 6 children were classified not at risk by the LLAS, yet had scores below the cut-off concern on the CDI Social (<0.8 quotient), one of whom has mild ASD (false negative on LLAS)
- 3 children in the study have been diagnosed with a form of ASD (2 with severe to profound HL), one of which was a false negative (did not screen positive) and the other two were noted as risk by criteria 3
- The LLAS may not be sensitive enough to pick out minute vocal qualities of children with milder forms of ASD

Perils in Diagnosing ASD in Deaf/HH

- “Gold standard” assessments/screening tools commonly used with hearing children have not been validated with children who are D/HH
 - ADOS, ADI-R, SCQ, MCHAT, SRS, GARS, CARS
- Use of ADOS-2 with D/HH
 - May under-identify ASD depending on how it is used
- Many tools may not reliably identify ASD among children who are D/HH
- Multiple sources of information and rule in/rule out process are necessary

Pearls for Diagnosing ASD in Deaf/HH aka: red flags

- **Atypical preverbal communication** (e.g., poor eye contact, lack of pointing, poor orientation for communication, poor joint attention) and delays in language acquisition beyond what one could expect based on hearing loss/etiology/intervention history
- **Atypical language features** (e.g., echolalia past phrase speech phase, palm rotation errors, persistent gesture use despite instruction in formal sign and use of formal sign by others in the child's environment (i.e., distinct from home signs))
- **Social difficulties** (.e.g, failure to initiate/respond to peers when communication taken into consideration, failure to recognize Deaf cultural norms, etc)
- **Repetitive behaviors/restricted interests** (visual inspection, etc)

Interventions for Dual Diagnosis

- Effectiveness of interventions for ASD and/or needed adaptations for interventional strategies severely lacking (evidence in the literature largely based on case studies).
- It is reasonable to take interventions which have been successful for hearing children to modify/adapt for children who are deaf/HH?

Review of Interventions for ASD

(Warren et al, 2011, Pediatrics)

- 4120 studies; 34 met inclusion criteria – 1 rated good – 10 fair – 23 poor
- Interventions thought to show improved outcomes in cognition, adaptive functioning & early educational attainment

Categories of Effective Intervention

(Warren et al., 2011, Pediatrics)

- Lovaas-based & Early Intensive Behavioral Intervention (EIBI)
 - Discrete trial teaching (DTT)
 - Widely known in the public as Applied Behavioral Analysis (ABA)
 - Uses praise & reinforcers → transfer to naturalized settings
- Comprehensive Approaches - Children < 2 yr
 - Early Start Denver Model → ABA techniques in a functional developmental framework, sensitive to developmental sequence, positive, affect-based relationship
 - 2 yrs enrolled – significant cognitive & language gains
 - Must be “implemented with fidelity” and supervision
- Parent Training
 - Best at promoting social communication & language; less impact on child’s IQ

Implications of Communication on Interventions for Dual Diagnosis

- Communication needed:
 - joint attention, turn-taking, imitation, choice-making, play
- Communication modality can be complex
 - Picture Exchange Communication System (PECS)
 - Technology/Augmentative Communication
 - Signs, gestures, spoken

Implementation of Interventions Children with ASD who are D/HH

- Lovaas/Early Intensive Behavioral Intervention
 - Direct teaching (breaking down a task and building the skill).
 - Generalization of skills learned
 - Finding appropriate motivators, rewards
- Comprehensive, developmental approaches
 - “What is ASD, what is hearing loss?”
 - Promoting interactions with typical peers – more challenges?
 - Begins early (12-18 mo.) – Delayed diagnosis of ASD in D/HH population may make this challenging?

Interventions for Dual Diagnosis: Social Communication

- Parent Training
 - Fostering social communication skills, teaching parents about importance of communication & language access in general
- Social Skills Groups
- Social Stories
- Who is the peer group?
 - Learning cultural norms for both hearing and Deaf worlds

Thoughts on Intervention in the Presence of Dual Diagnosis

- Don't give up on a child being able to use oral language/sign language necessarily without first targeting core symptoms of ASD (e.g., responsiveness to sound for kids with CI may be impacted by ASD, not indicator of failure of CI; difficulty acquiring sign language may reflect poor motor imitation skills common in ASD)
- Child may need different modalities for receptive & expressive language
- Individualize the intervention to address specific needs

Family Resources

- Gallaudet Odyssey special editions re: deafness/autism
 - www.gallaudet.edu/documents/clerc/odyssey-2008-v9i1.pdf and
www.gallaudet.edu/Images/Clerc/.../Odyssey_SPR_2012_Szymanski.pdf
- Deafness and Family Communication Center of the Department of Child and Adolescent Psychiatry- Children's Hospital of Philadelphia
<http://www.raisingdeafkids.org/special/autism/>
- Colorado Hands and Voices- Deaf Plus
<http://www.cohandsandvoices.org/plus/index.html>
- Autism Society <http://www.autism-society.org/>