

Medical and audiological management of young children dually diagnosed with hearing loss and an autism spectrum disorder

EHDI CONFERENCE 2018

Robyn Nolan, MD, Developmental Behavioral Pediatrician

Emily Nightengale, Au.D. CCC-A

Taylor Stevenson, Au.D. CCC-A

Debbie Mood, PhD, Licensed Clinical Psychologist



Children's Hospital Colorado
Here, it's different.™

Autism spectrum disorder in children with hearing loss:

- Reduced hearing usually identified first, delayed identification of ASD
- Early ASD symptoms may be mistake for symptoms of reduced hearing
- Behavioral challenges may make accurate audiology assessment difficult
- Additional medical problems that impact behavior can be missed while addressing larger diagnostic concerns



Etiology

- Genetics
 - ASD: 10-30% with underlying identifiable genetic etiology
 - Pre-lingual hearing loss: 70-80% with identifiable genetic etiology
- Environmental risk factors
 - Infections: congenital CMV, meningitis
 - Prematurity, low birth weight, cerebral palsy
- Teratogens: alcohol

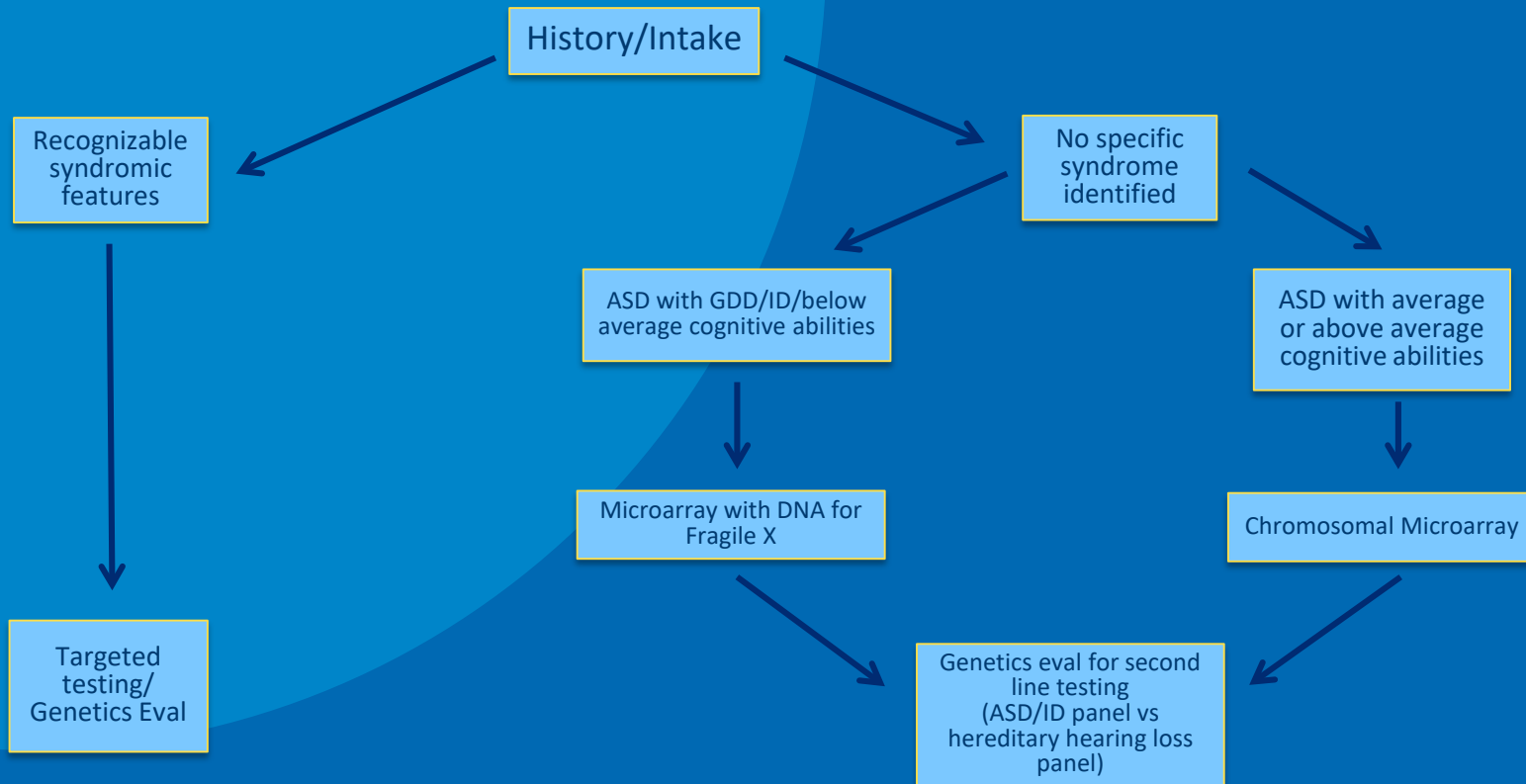


Examples of genetic overlap

- CHARGE
- Trisomy 21
- Usher Syndrome
- 22q11.2 deletion syndrome
- Goldenhar/Oculo-auricular vertebral syndrome



Genetic work up for ASD and hearing loss



Reasons for genetic testing

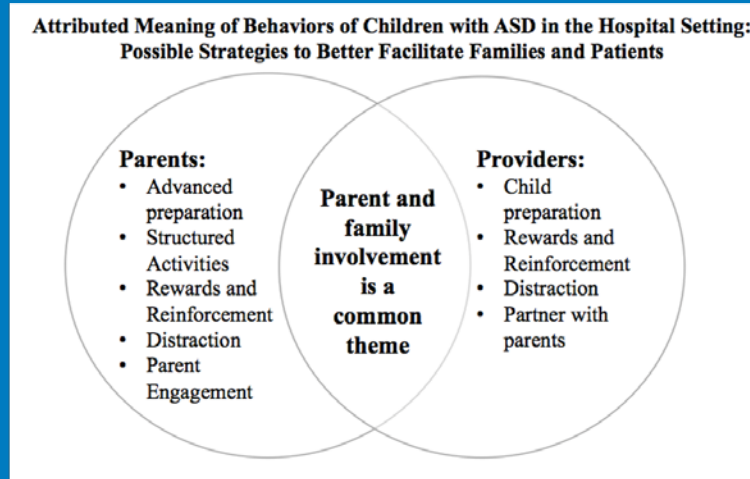
- Recurrence risk
- Understanding etiology
- Anticipatory guidance
 - Co-morbid medical conditions
 - Impact on recommended treatments



Autism and Audiology



Parent and Care Provider Perspectives



Focus group by Johnson et al. (2013):

- Health care providers reported attributing a child's challenging behaviors as self-stimulation and aggression
- Parents attributed this to child's communication frustration, hyperactivity, and self-calming



Autism and Test Performance

May not perform better (or worse) in the presence of an observer

- May not benefit from a human tester
- Perform at same levels with and without human testers
- *In fact:* in some environments, children with autism perform at a level comparable to typically developing peers when both use computer based programs, whereas typically developing peers perform better with human interaction and children with autism perform no differently

Need to better understand their “motivating factors”

- Intrinsic motivation is key to success: Special interests should be utilized
 - These interests should not necessarily be used as “reward”



Strategies for Testing:

How to prepare the soundbooth before child walks in:

- Ask about the child's interests/likes/dislikes
- Favorite toys, video, comfort items, sensory devices
- Remove distractions from booth
 - Eg. Cover computers/electronics with blanket
- Out of sight, out of mind
- Clear clutter - minimize distractions in room
- Show picture schedule again before you begin



Strategies for Testing:

What parents can do:

- Help child tolerate touch to head and ears

- Several times per day

- Head, shoulders, knees and toes

- Wear hats, earphones, headbands

- Playfully play with child's head/ears (massage)

- Use ear plugs at home to desensitize

- Visit clinic/booth prior to day of testing (take a tour)

- Show picture schedule (provided by audiologist)



Strategies for Testing

Difficulty shifting and dividing attention

- Can make timing of VRA challenging especially if child is more interested in something else in the room
- Inconsistent responses to environmental sounds

Combination of behavioral and objective measures likely necessary for children who have a higher tendency to have difficulty testing/ have higher false positive/false negative responses

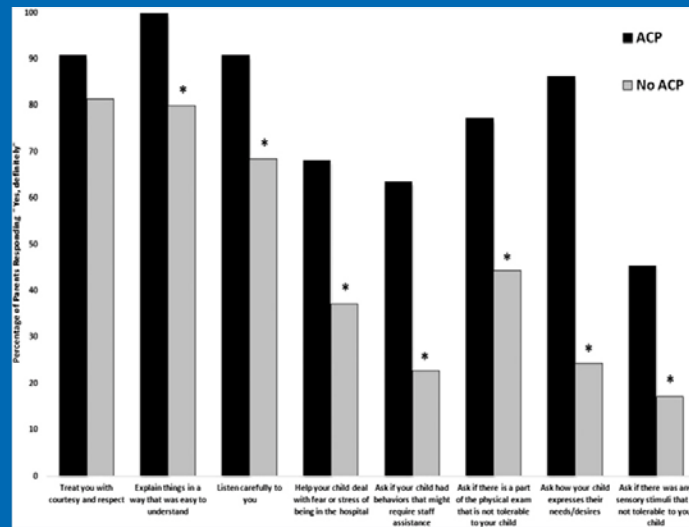
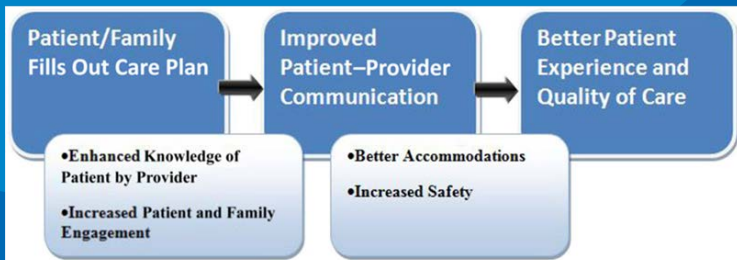
- May also need several visits
- Watch for atypical responses/behaviors/movements



Strategies for Testing: Adaptive Care Plans (prior to the appointment)

Types of questions that may be asked:

- How does your child respond to medical visits or hospital admissions?
- How does your child indicate that he/she is getting upset?
- What strategies have been previously used to manage behaviors that are a negative response to care or environment?
- What known anxieties or sensory sensitivities does your child have?
- What calming techniques does the child or family have?
- What is your child's preferred communication mode?



Use of care plans in a hospital setting
(parent satisfaction survey data)



A Family's Journey

Hearing loss diagnosis -

- Newborn hearing screen
- Diagnostic audiology evaluations
- Hearing aid fitting & earmold impressions

Early intervention -

- IFSP under part C of IDEA
- CHIP: Auditory training, auditory/verbal training, sign language, and verbal play
- Speech-language therapy & occupational therapy
- In transition to IEP under part B of IDEA with new providers and settings

ASD diagnosis -

- Developmental delays & suspicious behaviors
- Pediatrician referral
- Developmental pediatrician evaluation
- Therapy: Comprehensive behavioral therapy (ABA), communication, play skills, imitation, joint attention, self-help skills, and peer interaction



- Hearing Loss prevalence in ASD - No conclusive evidence to suggest there is a higher risk for hearing loss in ASD
- Sensory Processing disorder
 - Auditory defensiveness - Hyperacusis/Hyper-Responsiveness (18-53%); Serous Otitis Media (23.5%)
 - Behaviors include: crying, covering ears, vocalizing, avoidance, stiffness, even self injurious activities
 - Tactile defensiveness
 - Visual defensiveness



Children with ASD



Action Plan For Supporting Hearing Aid Use

- Child and Family Characteristics
- Outline potential challenges with possible solutions
- Preparing for use
 - Understanding child's current usage patterns
 - When to plan HA sessions
 - How to prepare child
- Initial goals and Rewards/Motivators
- Practice/Session guidelines



Hearing Device Multidisciplinary Support Clinic

Providers:

- Psychology and Audiology

Evaluation:

- History of Device Use
- Medical considerations
- Sensory considerations
- Developmental considerations
- Behavior
- Family Beliefs and Supports
- Observation
- Assessment Tools (i.e. Sensory Profile, Social Communication Questionnaire, Parenting Stress Index)

Treatment Plan and Recommendations:

- Body preparations
- Use of visual supports
- First/Then visual schedules
- Behavior management support groups
- Systematic desensitization program that inhibits anxiety and stress
- Sound desensitization hierarchy
- Desensitization games



Co-morbid medical conditions

Regression:

- Occurs in 30% of children with ASD between 1-3 years of age
- Change in speech can be seen with progression of hearing loss

Seizures

- 25-33% of children with ASD
- Can lead to regression or appearance of not responding well to sound



Co-Morbid medical conditions

Vision differences

- Reports of increased rates of strabismus, amblyopia, and refractive errors differences in ASD
- Vision differences associated with a number of syndromic causes of hearing loss

Motor/vestibular disorders

- Increased rates of motor impairments in ASD including motor planning, upper extremity motor control, and postural/balance concerns
- Increased risk of vestibular disorders in individuals with reduced hearing



Co-Morbid medical conditions

Sleep Problems

- 53-78% of children with ASD
- Increased risk in children with low vision or cortical vision impairment

Toileting/GI difficulties

- Increased rates of constipation and encopresis, diarrhea, bloating, toilet training delays, eosinophilic esophagitis in ASD
- GI symptoms associated with difficult behaviors



References

- Alford, R. L., Arnos, K. S., Fox, M., Lin, J. W., Palmer, C. G., Pandya, A., ... & Professional Practice and Guidelines Committee. (2014). American College of Medical Genetics and Genomics guideline for the clinical evaluation and etiologic diagnosis of hearing loss. *Genetics in medicine*, 16(4), 347.
- Beers, A., McBoyle, M., Kakande, E., Dar Santos, R., & Kozak, F. (2014). Autism and Peripheral Hearing Loss: A Systematic Review. *International Journal of Pediatric Otorhinolaryngology*: 78, 96-101
- Broder-Fingert et. al. (2016). A Pilot Study of Autism-Specific Care Plans During Hospital Admission. *Pediatrics*, 137.
- Chevallier, C., Parish-Morris, J., Tonge, N., Le, L., Miller, J., & Schultz, R. T. (2014). Susceptibility to the audience effect explains performance gap between children with and without autism in a theory of mind task. *Journal of Experimental Psychology: General*, 143(3), 972-979.
- Autism and Developmental Disabilities Monitoring Network Surveillance Year 2010 Principal Investigators. (2014). Prevalence of autism spectrum disorder among children aged 8 years—autism and developmental disabilities monitoring network, 11 sites, United States, 2010. *Morbidity and Mortality Weekly Report: Surveillance Summaries*, 63(2), 1-21.
- Dammeyer, J. (2012). Children with Usher syndrome: mental and behavioral disorders. *Behavioral and Brain Functions*, 8(1), 16.
- Fitzpatrick, E. M., Lambert, L., Whittingham, J., & Leblanc, E. (2014). Examination of characteristics and management of children with hearing loss and autism spectrum disorders. *International journal of audiology*, 53(9), 577-586.
- Gaffney, M., Eichwald, J., Grosse, S. D., & Mason, C. A. (2010). Identifying infants with hearing loss-United States, 1999-2007. *Morbidity and mortality weekly report*, 59(8), 220-223.
- Grove, R., Roth, I., & Hoekstra, R. A. (2015). The motivation for special interests in individuals with autism and controls: Development and validation of the special interest motivation scale. *Autism Research*, 9(6), 677-688.
- Hartshorne, T. S., Grialou, T. L., & Parker, K. R. (2005). Autistic-like behavior in CHARGE syndrome. *American Journal of Medical Genetics Part A*, 133(3), 257-261.
- Johnson, N. L., Bekhet, A., Robinson, K., & Rodriguez, D. (2014). Attributed Meanings and Strategies to Prevent Challenging Behaviors of Hospitalized Children With Autism: Two Perspectives. *Journal of Pediatric Health Care*, 28(5), 386-393.
- Kancherla, V., Braun, K. V. N., & Yeargin-Allsopp, M. (2013). Childhood vision impairment, hearing loss and co-occurring autism spectrum disorder. *Disability and health journal*, 6(4), 333-342.



References Continued

- Malow, B. A., Byars, K., Johnson, K., Weiss, S., Bernal, P., Goldman, S. E., ... & Glaze, D. G. (2012). A practice pathway for the identification, evaluation, and management of insomnia in children and adolescents with autism spectrum disorders. *Pediatrics*, 130(Supplement 2), S106-S124.
- Mannion, A., & Leader, G. (2014). Gastrointestinal symptoms in autism spectrum disorder: A literature review. *Review Journal of Autism and Developmental Disorders*, 1(1), 11-17.
- Miller, M. T., Strömmland, K., Ventura, L., Johansson, M., Bandim, J. M., & Gillberg, C. (2005). Autism associated with conditions characterized by developmental errors in early embryogenesis: a mini review. *International Journal of Developmental Neuroscience*, 23(2-3), 201-219.
- Myck-Wayne, J., Robinson, S., & Henson, E. (2011). Serving and Supporting Young Children with a Dual Diagnosis of Hearing Loss and Autism: The Stories of Four Families. *American Annals of the Deaf*: 156 (4).
- Niehus R, Lord C (2006). Early Medical History of Children with Autism Spectrum Disorders. *Developmental and Behavioral Pediatrics*. 27(2): S120-127
- Rosenhall, U., Nordin, V., Sandstrom, M., Ahlsen, G., & Gillberg, C. (1999). Autism and Hearing Loss. *Journal of Autism and Developmental Disorders*: 29 (5)
- Shearer, A. E., Hildebrand, M. S., & Smith, R. J. (2017). Hereditary hearing loss and deafness overview.
- Stiegler, L. & Davis, R. (2010). *Understanding Sound Sensitivity in Individuals with Autism Spectrum Disorders*. Focus on Autism and Other Developmental Disabilities: 25 (2) 67-75
- Szarkowski, A., Mood, D., Shield, A., Wiley, S., & Yoshinaga-Itano, C. (2014, November). A summary of current understanding regarding children with autism spectrum disorder who are deaf or hard of hearing. In *Seminars in speech and language* (Vol. 35, No. 04, pp. 241-259). Thieme Medical Publishers.
- Szymanski, C. A., Brice, P. J., Lam, K. H., & Hotto, S. A. (2012). Deaf children with autism spectrum disorders. *Journal of Autism and Developmental Disorders*, 42(10), 2027-2037.
- Thompson, D. G., & Tielsch-Goddard, A. (2014). Improving Management of Patients With Autism Spectrum Disorder Having Scheduled Surgery: Optimizing 20Practice. *Journal of Pediatric Health Care*, 28(5), 394-403.
- Warner, G., Howlin, P., Salomone, E., Moss, J., & Charman, T. (2017). Profiles of children with Down syndrome who meet screening criteria for autism spectrum disorder (ASD): a comparison with children diagnosed with ASD attending specialist schools. *Journal of Intellectual Disability Research*, 61(1), 75-82.

