LEND Pediatric Audiology Training Program
Pre-EHDI Workshop
April 13, 2014

Allan's Story: A Case of Autism Spectrum Disorder and Hearing Loss by Vicki Simonsmeier

Learning Objectives:
At the conclusion of this story, participants will be able to:

- Understand the effects of hearing loss and ASD on development;
- Recognize the various disciplines involved in making the ASD & hearing loss diagnoses;
- Define components of the best practices approach to diagnosis of autism;
- Refer families to appropriate resources.
- Add more objectives

Part I: The Dell's

Allan, the son of Amy and Robert Dell, is a 5 year, 2 month old right-handed Caucasian male who was referred to the Autism Spectrum Disorders Clinic because of concerns about a possible autism spectrum disorder. Allan has a history of moderate to severe bilateral sensorineural hearing loss first identified at age 18 months for which he has received early intervention services at a program for children with hearing loss. Referral questions from Mr. & Mrs. Dell state that Allan displays delays in social behaviors such as lack of eye contact, lack of parallel play, and lack of interaction with peers. There are also concerns with sensory issues and he has been slow to progress in multiple areas in school. There also are concerns about developmental delays.

The initial interview elicited the following information from Mr. & Mrs. Dell. Allan was developing normally up until 13 months of age. He was able to sit at 6 months, crawl at 7 months, and was starting to talk by 12 months, including phrases ("more milk please"). He had problems with multiple respiratory infections starting about 10 months of age. At 13 months of age he received 3 vaccines, MMR, HIB, and Hep B. He was recovering from a respiratory infection at the time of vaccination. Two hours after receiving the vaccines, he developed a fever (104.2 degrees), was inconsolable, had no evident seizure, but appeared to be tremoring or shivering. Mr. & Mrs. Dell alternated Tylenol and ibuprofen. Allan also developed a sandpaper type rash, and was very pale. The rash persisted for about 3 weeks. He was ill off and on for the next 3 months, with reported 19 medical office visits during that time. His parents initially attributed his developmental lag at that time to being sick so often. Allan referred on the otoacoustic emissions (OAE) screen at at birth but passed auditory brainstem response (ABR) screening at 3 weeks of age. Following the illnesses and respiratory problems, Allan was evaluated when he was 18 months by Dr. Low, ENT, who diagnosed obstructive sleep apnea. He underwent tonsillectomy/adenoidectomy and ventilation tube placement. It was at this time Allan was found to have a moderate to severe bilateral, sensorineural hearing loss. He was fit with hearing aids "with good effect". Allan was referred for Connexin 26 genetic workup. Ultimately these tests were negative for Connexin, Fragile X, nor any ASD related disorders.

1. *What additional information would you like about Allan and his family?*
2. *What counsel could you provide parents regarding their concerns that Allan's vaccinations caused his hearing loss?*
3. *Given what you know thus far about Allan and the parental concerns, what other areas of assessment might be necessary?*
4. *How would you explain to parents that Allan's hearing assessment results at birth fit or do not fit with the audiological findings from 18 months of age?*

Part II: Additional assessment data

As part of the autism diagnostic evaluation, the team completed a detailed review of Allan's records with the following information summarized below.

At age 3 years 7 months, Allan was seen for a consideration of an autism spectrum disorder. At this time there were concerns about lack of eye contact, sensory issues, and a lack of parallel play. The physician noted a moderate to severe hearing impairment and suspicions of a global developmental delay and perhaps not an autism spectrum disorder. Recommendation was for additional assessment by a psychologist to determine Allan's level of functioning.

At age 3 years, 10 months, Allan was referred by his preschool program for evaluation of possible ASD. On the Stanford-Binet Intelligence Scale, 4th Ed., Allan obtained scores in the extremely low to borderline range, with a total test battery score of 69 (100 equals average,+/-15). It was noted that the test scores should be interpreted cautiously as Allan displayed difficulties with receptive and expressive language and difficulty manipulating some of the test materials. The examiner stated "present scores are significantly underestimating his true overall abilities, and he does present like a child whose overall level of intellectual functioning seems to be more within the average range." He also was administered the Autism Diagnostic Observation Scale (ADOS). Allan was diagnosed with a Mixed Language Delay, Speech Delay, Fine Motor Coordination Problems, Sensory Integration Difficulties, and a provisional diagnosis of Pervasive Developmental Disorder Not Otherwise Specified.

In the fall at 4 years, 7 months, Allan was evaluated at his preschool. On the Peabody Picture Vocabulary Tests, 4th Edition, a measure of receptive vocabulary, Allan obtained a standard score of 58 (average standard score = 100 ± 15), which was well below average. On the Clinical Evaluation of Language Fundamentals, Preschool Test, administered September 2011, he obtained overall scores that "places Allan in the very low-severe range when compared to same-age peers."

At age 4 years, 10 months, the preschool had concerns with Allan's behavior in the classroom and during therapy activities. Allan demonstrated multiple developmental problems including language delay, social pragmatic delays, and delays in fine and gross motor skills. It also was noted that Allan "does not engage in interactive play skills consistent with his chronological age peers but rather manifests parallel play behaviors." It was documented that Allan was displaying more and more off-task behaviors, and that his parents had reported more inappropriate behavior at home. The off-task behaviors included lying on the floor, refusing to take turns in classroom routines, touching and feeling objects in an apparent sensory-seeking manner, and verbal protests and refusal of activities and not following directions. Specific behaviors included turning away, screaming, pushing peers, and removing his hearing aids. Allan also had been displaying a lack of eye contact and not engaging in interactions with others, intruding on the personal space of others, and oral stimulation such as chewing his shirt and licking objects. He also had begun to display self-injurious behaviors such as incurring carpet burns by rubbing his head on the carpet excessively. Finally it was noted that Allan has demonstrated slow progress towards his speech and language goals, struggling with two-item memory tasks, spatial concepts, and speech production targets.

At age 5 years, 1 month a psychoeducational evaluation documented that the Scales of Independent Behavior – Revised were administered to Allan's mother. This is a measure of adaptive behavior and it was noted "functional independence is limited" with an age-equivalent score of 2 years, 7 months. On the Behavior Assessment System for Children – II, a standardized behavior checklist completed by his mother, ratings were significant for hyperactivity, anxiety, atypical behavior, and withdrawal. Allan was also seen for a for a neuropsychological evaluation. Dr. Sky documented his developmental history and concerns about "the complexity of his developmental challenges and in preparation for a possible cochlear implant." On the

Differential Abilities Scale, 2nd Ed., a standardized measure of cognitive skills, Allan obtained a General Conceptual Ability score of 47 (average is 100, ± 15). Dr. Sky concluded that Allan presented with symptoms and behaviors consistent with a neurologically-based cognitive impairment similar to that of children with a traumatic brain injury, and "as such he cannot be viewed as a child with only a hearing impairment." She diagnosed Allan with a Cognitive Disorder Not Otherwise Specified, and Pervasive Developmental Disorder Not Otherwise Specified.

In the "Team Evaluation Summary Report and Prior Notice of Eligibility Determination: Developmental Delay" when Allan was 5 years of age, it was recorded that Allan met criteria a classification of Developmental Delay.

1. *What are some red flags for autism spectrum disorders? Does Allan exhibit any of these factors?*
2. *Given the totality of these tests, have the parents original questions been answered (see referral question in Part I), why or why not?*
3. *Do you feel that these diagnoses fully define Allan's abilities, why or why not?*
4. *Has this family received care consistent with the MCH guidelines of family centered, community based, culturally competent, interdisciplinary care? What components of the MCH guidelines is missing and how might this have advanced care for this child?*

Part III: Interdisciplinary Evaluation

At age 5 years, 2 months Allan saw an interdisciplinary team (medical, audiological, psychological, occupational therapy, speech-language pathology) who made the following diagnoses:

Speech//Language Evaluation
Allan meets criteria for severe mixed, receptive and expressive language disorder. Standard test scores were in the 50's for receptive and expressive language, with age equivalent scores in the 30-36 month range. On a criterion-referenced social-pragmatic assessment, Aidan demonstrated strengths in the areas of imitation of actions, turn-taking, eye gaze, joint attention, and the ability to transition quickly between activities. Aidan readily imitated actions performed by the clinician, and his mother (e.g. stacking blocks, blowing bubbles). Aidan demonstrated difficulties in the areas of symbolic play, even with a great deal of modeling it was difficult to elicit symbolic play from Aidan.

Audiology Evaluation
Bilateral, moderate to severe sensorineural hearing loss, with adequate hearing levels achieved with amplification.

Psychology Evaluation
Allan meets criteria for Cognitive Disorder Not Otherwise Specified (DSM-VI-TR). Cognitive performance on the Leiter International Performance Scale Revised revealed a Brief IQ of 73 (standard score).

Occupational Therapy
Allan was also diagnosed with a sensory processing disorder. Motor Coordination disorder was also diagnosed.

Developmental Pediatrician
Allan meets criteria for Pervasive Developmental Disorder Not Otherwise Specified according to the American Psychiatric Association's Diagnostic and Statistical Manual of Mental Disorders (DSM-IV-TR). This included information from the ADOS, the ADI-R, parent report checklist and behavioral observations in the child's school. A medical recommendation for an EEG was made to rule-out seizure disorder.

1. *Considering the diagnoses listed above, is this new information for the parents, why or why not?*
2. *Why wasn't Allan making progress in preschool commensurate with his peers?*
3. *Why has this family been asking these same questions for the past 4 years? What factors may have played a role in the odyssey this family has been on for 4 years?*
4. *What resources are available to this family? What treatment or educational options are appropriate?*

Part IV: Conclusion?