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Early Intervention for Autism Spectrum Disorder in Children who are Deaf/Hard of Hearing: Identification: Part 1-Screening

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Children's Hospital Colorado
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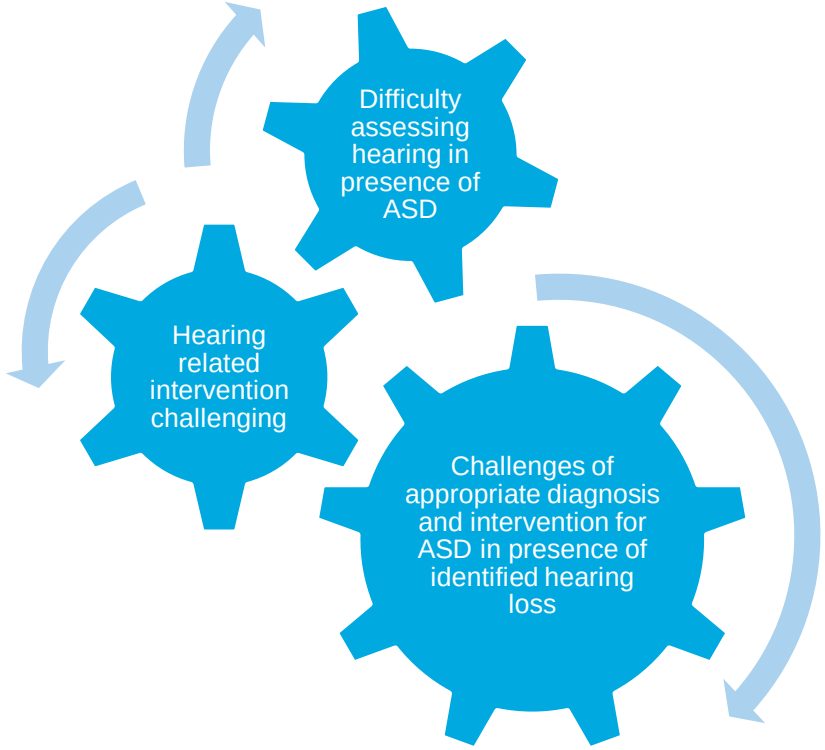
OBJECTIVES

1. Learners will describe “red flags” for ASD among children with reduced hearing.
2. Attendees will recognize the unique ways that ASD can present among children who primarily communicate using sign language.
3. Describe strategies for sharing concerns with families and facilitating appropriate referrals.

Hearing loss = diagnostic rule out + complex comorbidity



Complex comorbidity and complex differential diagnosis

Pediatric hearing loss-

1 to 3: 1,000- newborn

5 :1,000 children ages 3-17

Higher risk for hearing loss:

-prematurity

-NICU stay (2-4:100)

-ototoxic medications

-family history

-infections (CMV, meningitis)

-NICU stay with ECMO

-underlying genetic conditions

DSM-V criteria of ASD

Must meet criteria A, B, C, and D:

A. Persistent deficits in **social communication and social interaction** across contexts, not accounted for by general developmental delays, and manifest by all 3 of the following:

1. Deficits in **social-emotional reciprocity**; ranging from abnormal social approach and failure of normal back and forth conversation through reduced sharing of interests, emotions, and affect and response to total lack of initiation of social interaction,

2. Deficits in **nonverbal communicative behaviors used for social interaction**; ranging from poorly integrated- verbal and nonverbal communication, through abnormalities in eye contact and body-language, or deficits in understanding and use of nonverbal communication, to total lack of facial expression or gestures.

3. **Deficits in developing and maintaining relationships**, appropriate to developmental level (beyond those with caregivers); ranging from difficulties adjusting behavior to suit different social contexts through difficulties in sharing imaginative play and in making friends to an apparent absence of interest in people

DSM-V continued

B. **Restricted, repetitive patterns of behavior**, interests, or activities as manifested by at least two of the following:

1. **Stereotyped or repetitive speech**, motor movements, or use of objects; (such as simple motor stereotypies, echolalia, repetitive use of objects, or idiosyncratic phrases).

2. **Excessive adherence to routines**, ritualized patterns of verbal or nonverbal behavior, or excessive resistance to change; (such as motoric rituals, insistence on same route or food, repetitive questioning or extreme distress at small changes).

3. **Highly restricted, fixated interests** that are abnormal in intensity or focus; (such as strong attachment to or preoccupation with unusual objects, excessively circumscribed or perseverative interests).

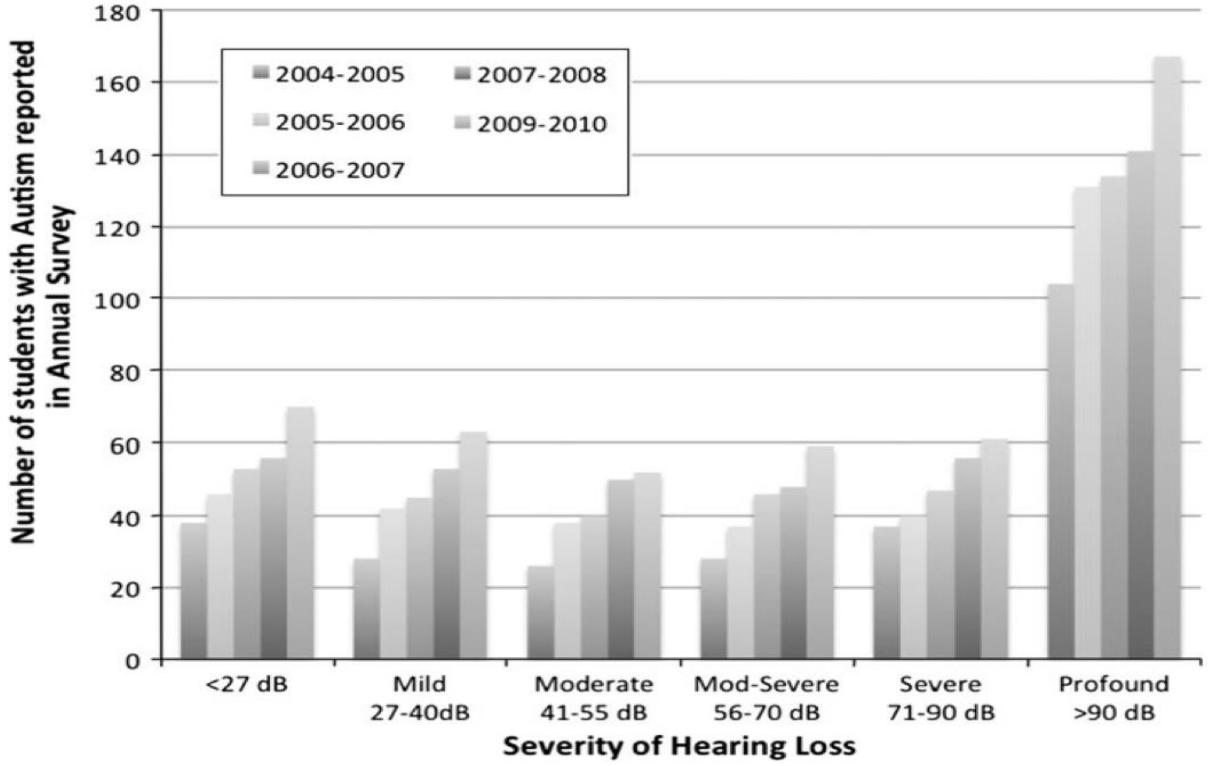
4. Hyper-or hypo-reactivity to **sensory input** or unusual interest in sensory aspects of environment; (such as apparent indifference to pain/heat/cold, adverse response to specific sounds or textures, excessive smelling or touching of objects, fascination with lights or spinning objects).

DSM-V

C. Symptoms must be present in early childhood (but may not become fully manifest until social demands exceed limited capacities)

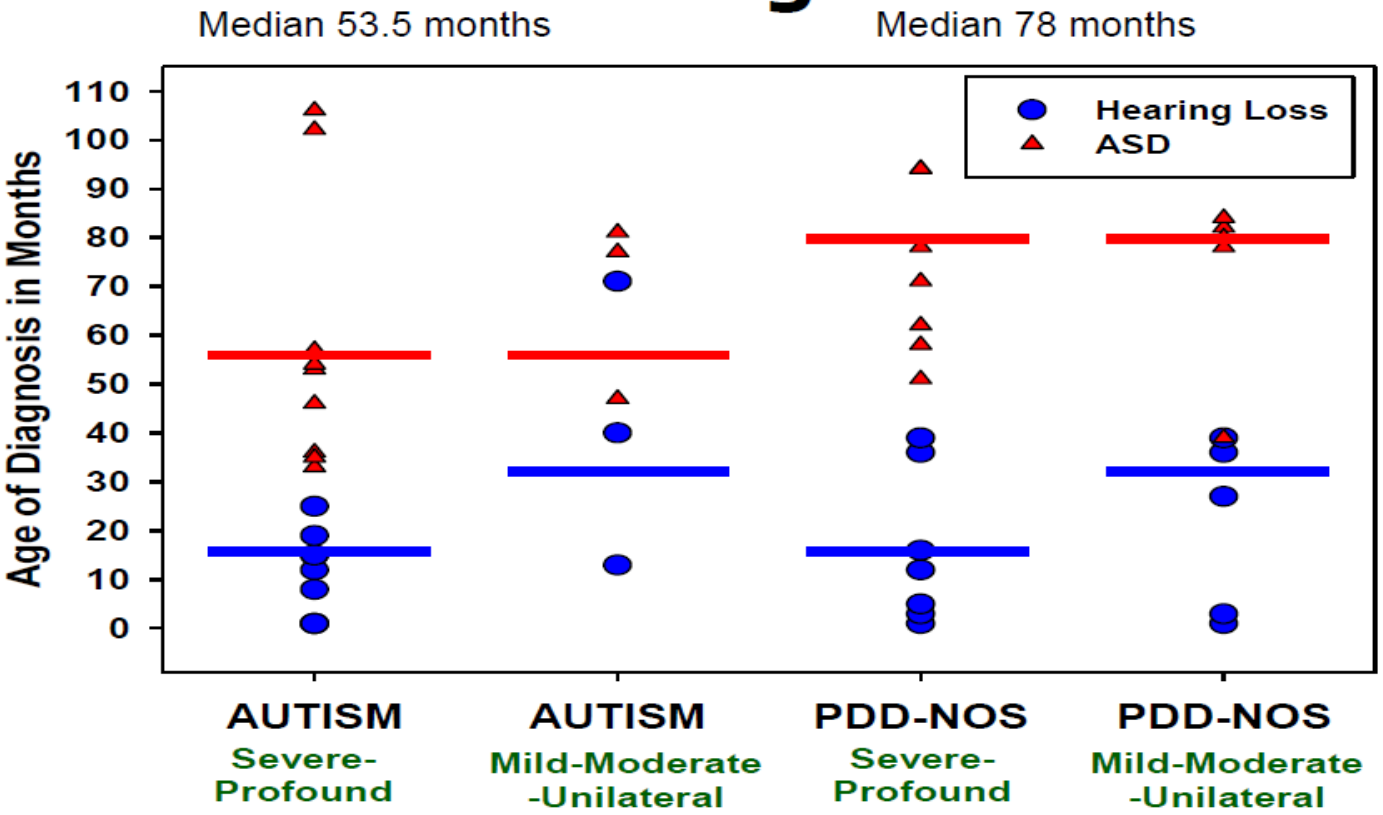
D. Symptoms together limit and impair everyday functioning.

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

Age of Diagnosis for ASD and Hearing Loss



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

Which children who are D/HH are most at-risk?

- Symptomatic CMV
- CHARGE syndrome (?)
- Prematurity
- Usher syndrome (?)
- Down syndrome

Syndromic causes of hearing loss with developmental consequences

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

Earlier identification

→ earlier intervention

→ improved outcomes

ASD can be reliably diagnosed ~ 18 months

Many possible “red flags” emerge ~ 12-15 months

Early Reduced Social Communication Skills

- Poor eye contact
- Lack of posturing for social communication
- Reduced social smile
- Reduced showing

Additional Early Social Communication Challenges

- Reduced giving for shared enjoyment
- Poor joint attention
- Reduced imitation
- Reduced gesture use

Developmental Nature

- Symptoms qualitatively different at different stages of development
- Negative symptoms (e.g., lack of imitation at 18 months) may be more indicative of autism than positive symptom approach because many of the positive symptoms are often present in other neurodevelopmental disabilities

Early vs Later

- Look for what's missing
- Consider ***quality*** and ***function*** of social/communication
  - To direct and regulate
  - To request vs to share
  - Engagement and play

- Presence of atypical communication
  - Echolalia
  - Stereotyped language
  - Pronoun confusion
  - Atypical prosody
  - Idiosyncratic language

Regression

- Loss of previously mastered skills warrants referral to PCP and/or for evaluation
  - May also warrant updated audiological evaluation
- Distinguishing from hearing change
  - Typical ASD regression between ages 12-24 months
  - Decline in speech/language only or also other skills
  - Return to baseline functioning

Delays in Language Warrant an Evaluation

-Anything beyond what one could expect based on hearing status, etiology, or intervention history

- Consider the “why”

“A lack of progress should never be assumed to be only due to a mismatch of chosen communication modality or insufficiencies in the language stimulation environment”

(Wiley & Innis, 2014)

- Although not a “core symptom”, language deficits go hand-in-hand with ASD
  - True for deaf/HH as well as hearing
- Shield study of Deaf of Deaf children with ASD
  - About 1/3 had minimally expressive sign
  - On average, receptive ASL skills were significantly below peers
- Suggests that exposure to sign in and of itself is unlikely to be a “silver bullet” in the case of ASD even though often used as an intervention for hearing children with ASD
- May need alternative communication strategies such as AAC

Characteristics of ASD language features in sign and speech

Similarities	Differences
Jargon	Pronoun reversal (speech only?)
Echolalia	Palm reversals (sign only)
Pronoun avoidance/Use of names	Facial grammar - impaired?
Lack of expressive language Deficits in receptive language	Spatial grammar - classifiers, agreement verbs
Idiosyncratic language	
Pragmatic deficits	

SHIELD (2014), SEMINARS IN SPEECH & LANGUAGE

- Palm reversals during signing and imitating
- Appears to reflect a different approach to imitation: ***imitate exactly what you see***

SHIELD & MEIER (2012), JOURNAL OF COMMUNICATION DISORDERS

- **Diagnosis and assessment**:
  - Unique error type could be a red flag for deaf children (beyond age 2)
- **Intervention:**
  - Signing next to a child with ASD may be more effective than face-to-face

A Deaf mother signing in front of her child

Echolalia in ASL

SHIELD, COOLEY, & MEIER (IN PREP)

Click to edit Master title style

Social communication

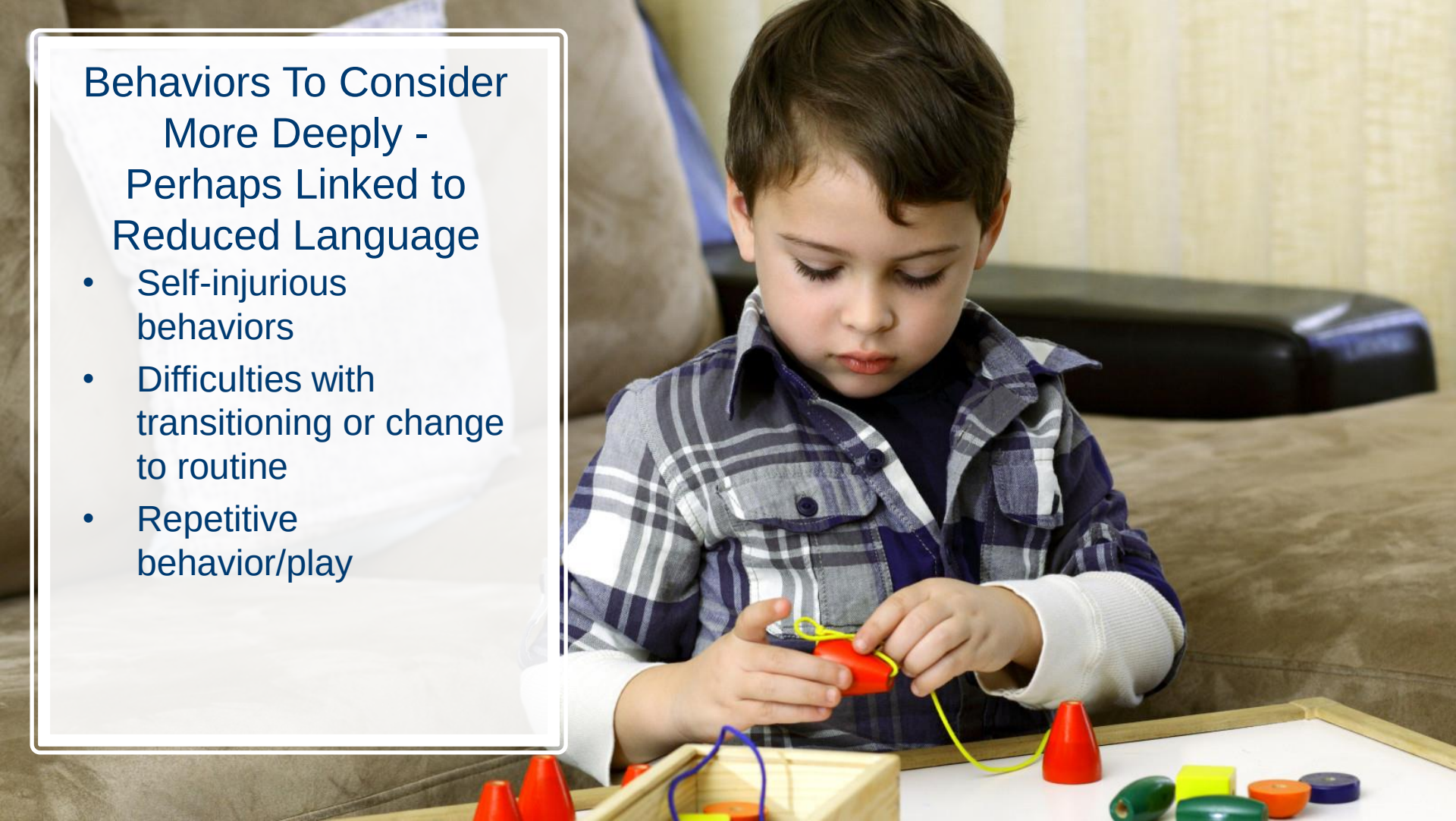
-Symptoms must be considered within context of communication access *and* cultural context

-Early pre-verbal social communication skills should not be impacted

-Social interactions mediated by language vs. less language demand

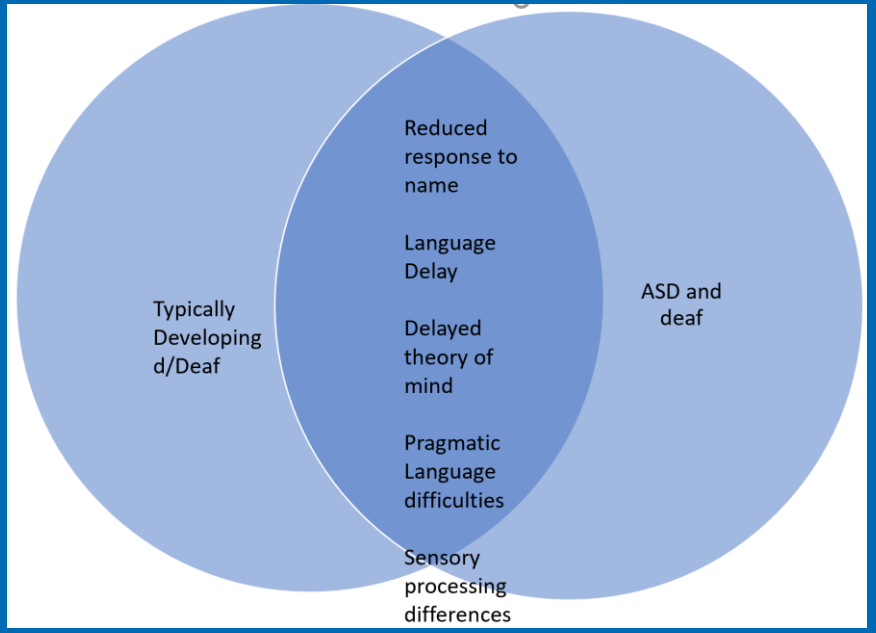
Repetitive & Restricted Behaviors

- Intense/unusual interests
- Repetitive body movements
- Unusual sensory exploration
- Sensory processing difficulties



Behaviors To Consider More Deeply - Perhaps Linked to Reduced Language

- Self-injurious behaviors
- Difficulties with transitioning or change to routine
- Repetitive behavior/play



Differential Diagnosis

Differential diagnosis requires understanding impact of language access

Developmental stages of acquiring sign language are still being defined

Differentiating “language deprivation” from symptoms of ASD

Pragmatic language skills

- Goberis, Beams and Yoshinaga (2012)
- Wiley et al (2017) “language underperformers”

Factors influencing differential diagnosis

- Etiology of hearing loss
- Age of identification
- Intervention history
- Consistency of use of hearing technology
- Communication access
- Access to peers
- Vision
- Vestibular concerns

Screening tools for ASD

1. Modified Checklist for Autism in Toddlers, Revised (MCHAT-R)
2. Screening Tool for Autism in Toddlers and Young Children (STAT)
3. LENA
4. Social Communication Questionnaire (SCQ)
5. Social Responsiveness Scale- Second Edition (SRS-2)

Screening children who are deaf or hard of hearing for autism

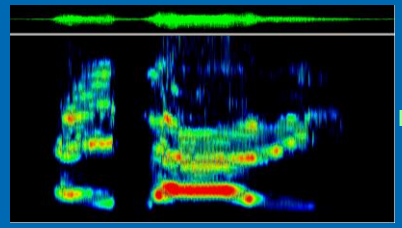
LENA: Language ENvironment Analysis

LENA Methods: Automatic Processing

Audio Stream of Child Voice
& Environment Sound

Identification of Different Sounds (Segmentation)

Sequence of
Key Child,
Adult,
Environment Noise
Overlapped Sounds
.....



Human Voice
(Child or Adult)

Phone Recognition

Consonant-like Sound,
Vowel-like Sound,
Non-Speech Sound,
Pause

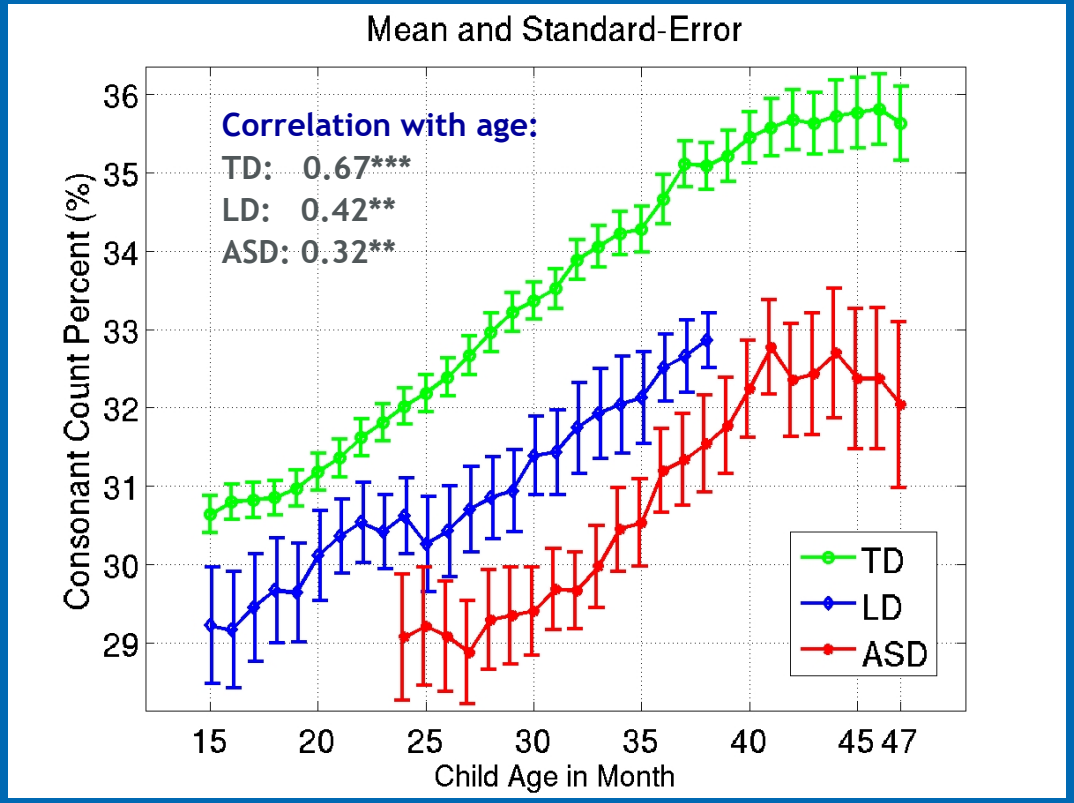
Data Set of the Study

Child Groups	Number of Children (N)	Number of Recordings	Child Segments (number in million)	Phoneme-like Units (number in million)
Typical Development (TD)	106	802	2.15 M	8.42 M
Language Delay but not ASD (LD)	49	333	0.75 M	2.65 M
Autism (ASD)	71	225	0.53 M	1.82 M
Total	226	1363	3.43 M	12.89 M

In the following slides of results of findings

- Green: Typical Development (TD)
- Blue: Language Delay not Related to Autism (LD)
- Red: Autism (ASD)

Frequency of Consonant-like Sound



t-test
(Welch 2-sample 2-side)

TD versus ASD:
t(90) = 7.95***

TD versus LD:
t(68) = 5.52***

LD versus ASD:
t(118) = 2.62**

*p<0.05
**p<0.01
***p<0.001

Correlation with chronological -age:

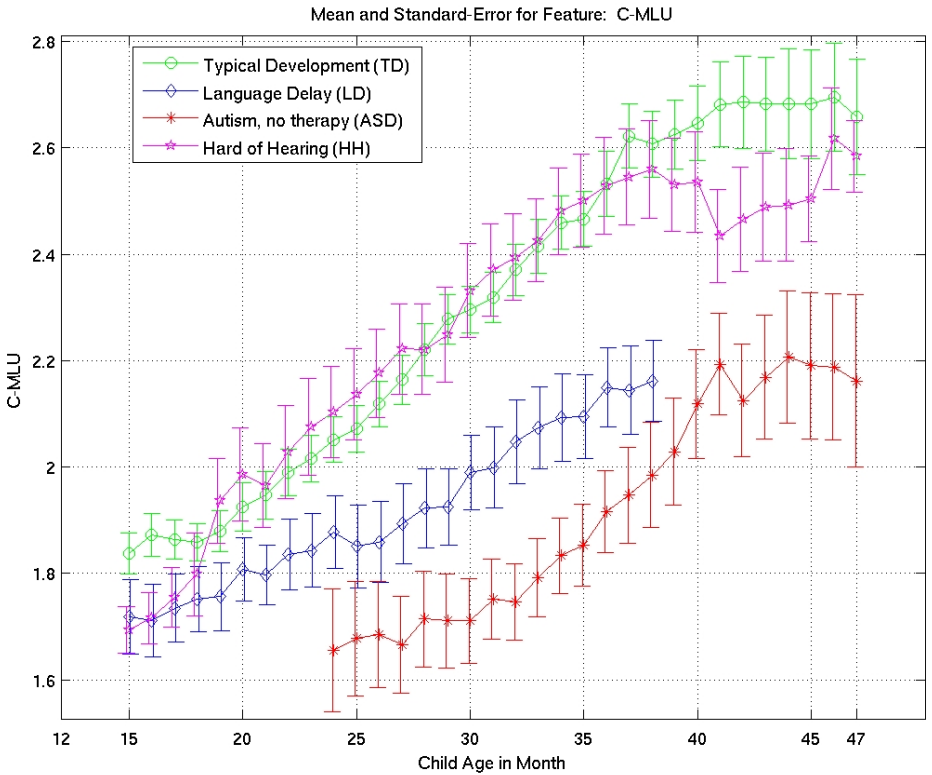
HH: 0.51 ***

TD: 0.63 ***

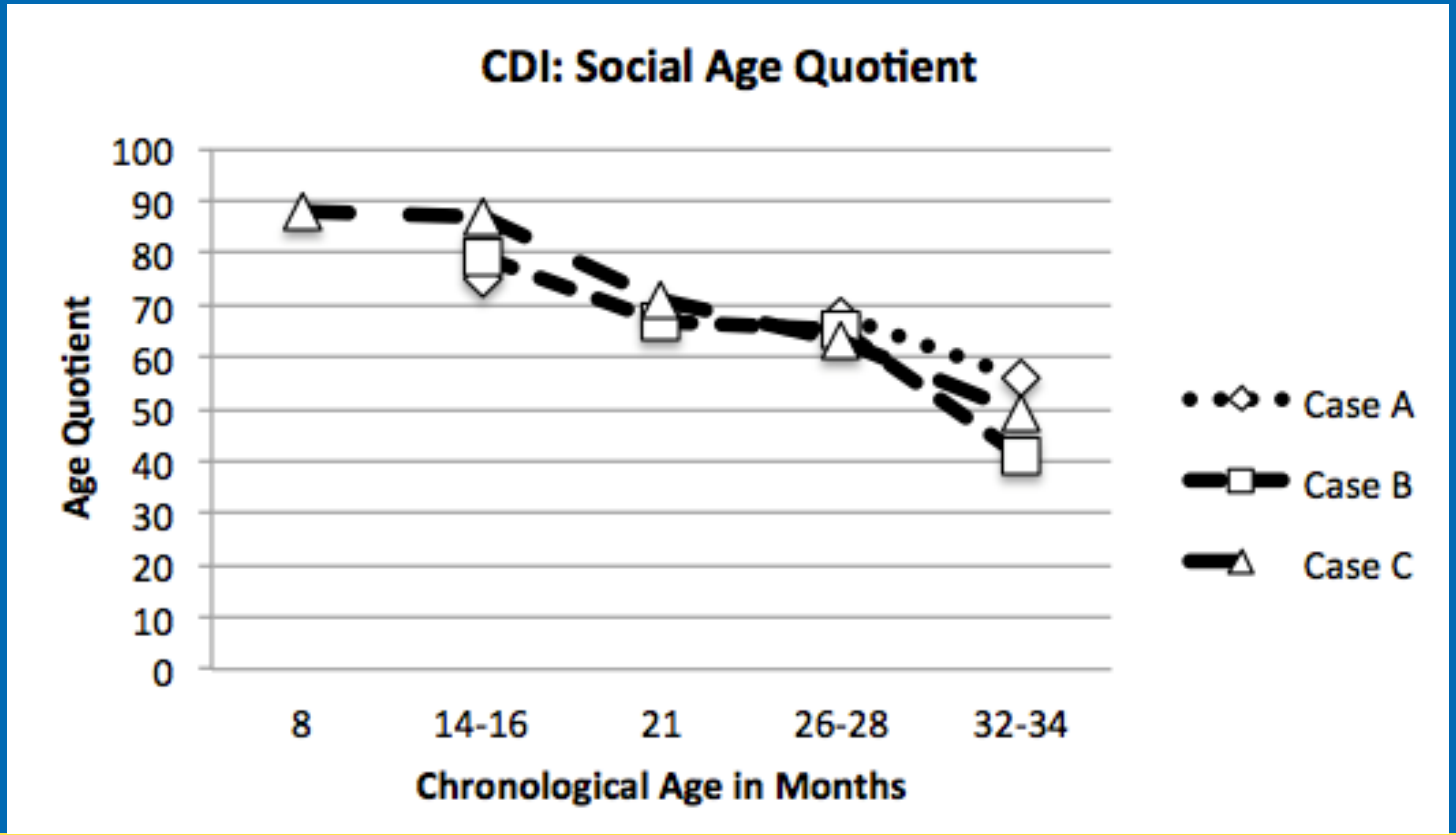
LD: 0.32 *

ASD: 0.32 *

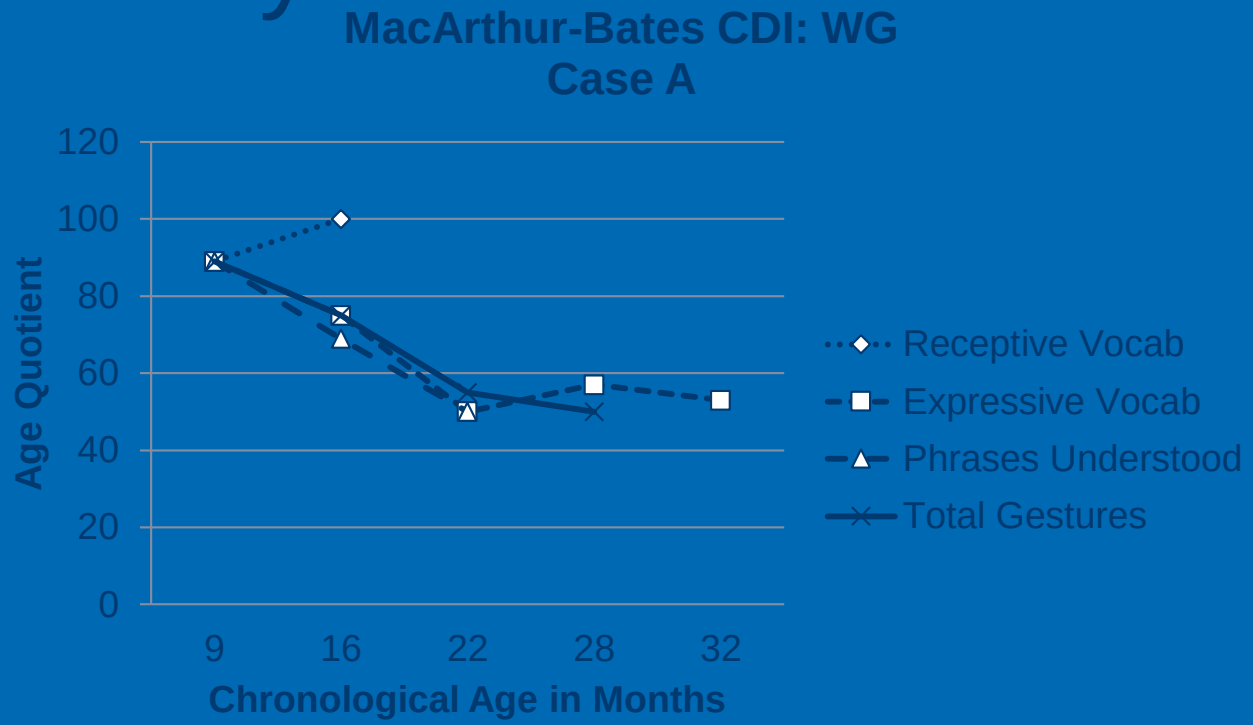
*: p < 0.05
**: p < 0.01
***: p < 0.001

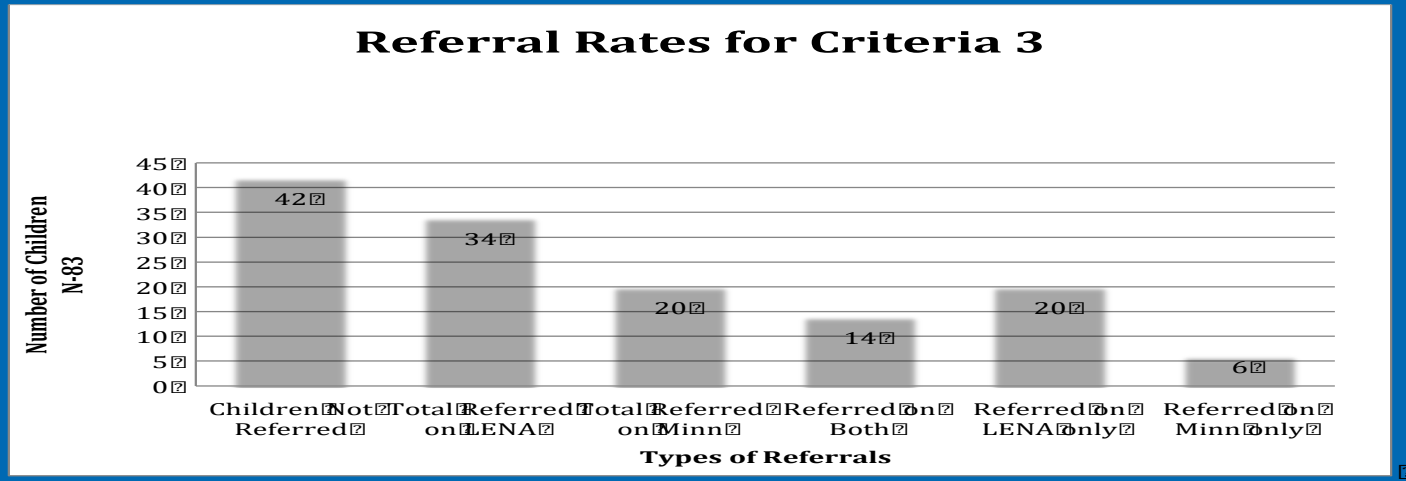


LENA + Child Development Inventory Social Age



MacArthur-Bates CDI: Symbolic Gesture




Referral Rates for Criteria 3
Number of Children
N-83
45
40
35
30
25
20
15
10
5
0
42
34
20
14
20
6
Children Not Referred
Total Referred on LENA
Total Referred on Minn
Referred on Both
Referred on LENA only
Referred on Minn only
Types of Referrals

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

LENA™ Language and Autism Screen (LLAS)

Accuracy for ASD Screens vs. LLAS

Screen name	N	Age	Administration	Time	Sensitivity	Specificity
LLAS – 1st Recording (24-48 mo. sample)	190 (75 ASD)	24-48	AUTOMATIC	–	.89	.89
CHAT (Checklist for Autism in Toddlers)	16,235 (12 ASD)	Avg.=19 mo.	Parent report 14 items; clinical observation	5-10 min.	.18	1.0 (100%)
M-CHAT (Modified Checklist for Autism in Toddlers)	1,293 (39 ASD)	18-30 mo.	Parent report 23 items	3 min.	.87	.99
ESAT (Early Screening for Autistic Traits)	31,724 (18 ASD)	13-23 mo.	Parent report 14 items	5 min	.90	.99
ABC (Autism Behavior Checklist)	133 (38 ASD)	4 -11 years	Caregiver checklist of 57 behaviors	5-10 min	.92	.93
ASQ (Autism Screening Questionnaire) now called the SCQ	200 (160 ASD)	4-40 years	Clinician checklist of 40 questions	5-10 min	.85	.75

Parent Measures

- Social Communication Questionnaire
  - Children over age 4; mental age over age 2
  - Cut off over 15= referral for full evaluation
  - English and Spanish version
- Social Responsiveness Scale- Second Edition
  - Ages 2 through adulthood
  - Parent and teacher rating scales
  - Rates Domains: Social Awareness, Social Cognition, Social Communication, Social Motivation, Restricted Interests and Repetitive behavior
- Autism Diagnostic Interview- Revised
  - Children and adults with mental age >2
  - Standardized interview yields scores mapped onto algorithm
  - English and Spanish version

LEVEL II

- Direct Assessment Instruments
  - Autism Diagnostic Observation Schedule (ADOS-2)
  - Childhood Autism Rating Scale (CARS)
  - Gilliam Autism Rating Scales (GARS)
- “Gold Standard”
  - ADI-R or similar interview
  - ADOS-2
  - Clinician’s Judgment

"Gold standard" assessment tools commonly used with hearing children have not been validated with children who are D/HH

- Efforts underway in Great Britain to validate ADOS-2, SRS-2, SCQ for use with D/HH

Use of ADOS-2 with D/HH (Mood & Shield, 2014)

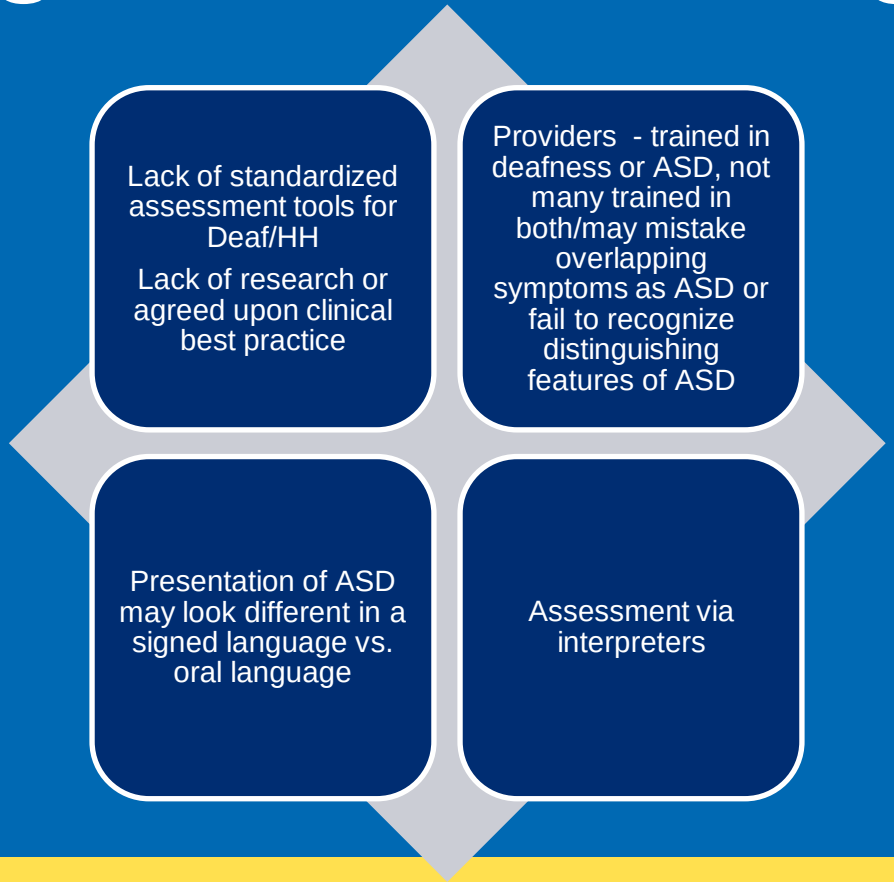
- May under-identify ASD if used in a "standardized" manner
- Failure to administer module that matches the child's language functioning results in lack of ability to assess atypical language and social communication
- Administration of "easier" module relies on tasks that are too developmentally easy and a missed opportunity to assess social/communication skills appropriate for the child's developmental functioning

Many tools may not reliably identify ASD among children who are D/HH

- Use of ADOS-2 algorithms with D/HH is not advised
- Screening tools such as MCHAT-R and SCQ can both over and under-identify D/HH children with ASD
- Advisable to still refer a child who presents with red flags for ASD who "passes" these screening measures for a more comprehensive assessment
- When used by a clinician familiar with ASD and deafness, ADOS-2 may reveal important clinical information

Multiple sources of information and rule in/rule out process are necessary

Diagnostic Challenges



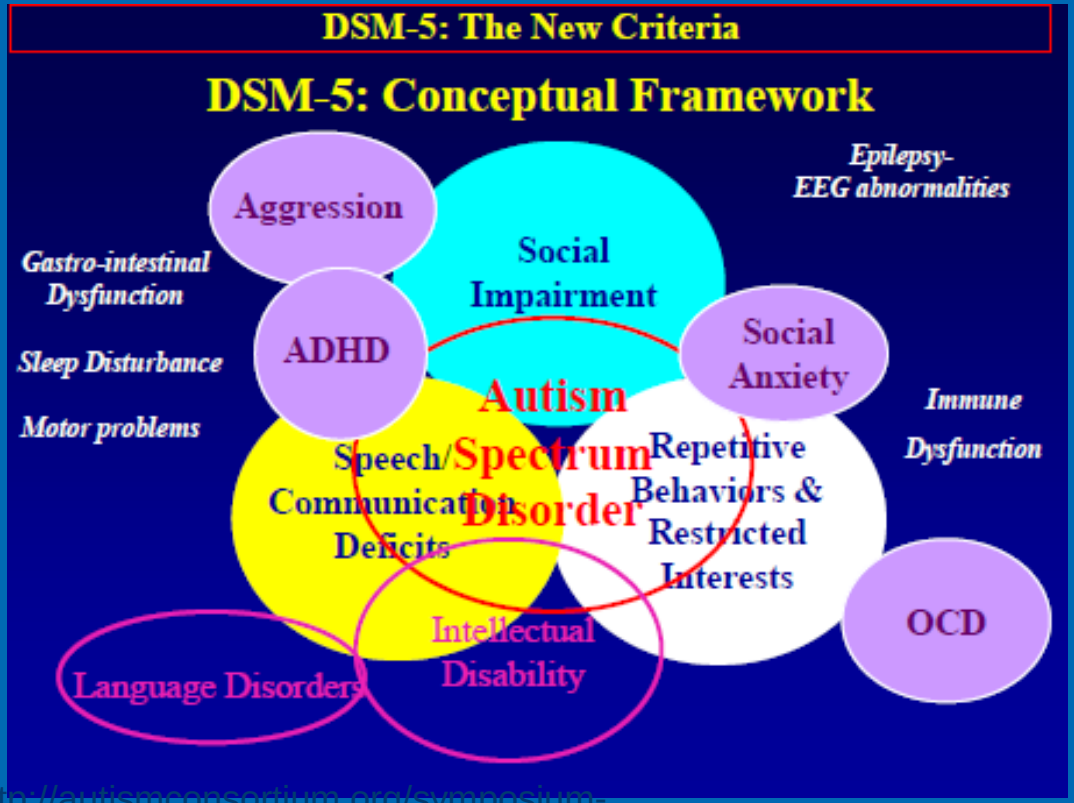
Earlier identification = earlier treatment = better outcomes

- Audiologists, TOD, SLPs, early interventionists often first to recognize symptoms which are not typical for hearing loss
- Key role in facilitating early identification and earlier intervention

Why refer?/ Why diagnose?

- Differentiated treatment
- Educational placement
- Communication decisions
- Financial resources
- Parental validation
- Identity
- Medical

The “Big” picture



http://autismconsortium.org/symposium-files/WalterKaufmannAC2012Symposium.pdf

Sharing Diagnostic Information with Families

"Wait & See"

Parental discontent

Resentment & anger

Carbone, 2012

Supporting Families

Do not delay raising concerns with families

Families may not understand what is/is not attributable to hearing loss

Start with strengths and discuss concerns in a kind, clear manner

Help families recognize symptoms of ASD are broad: filter what does/does not apply to their child

Be able to help families identify resources for seeking diagnostic clarification and support

Connect with other families with children with similar strengths/needs

Joining with families

- Discuss observations without judgment, but with professional guidance, providing context for observed strengths and needs
  - Did you notice? … I would typically expect…. That behavior is different than what we typically expect of a child with x hearing loss
  - Explore family concerns/fears
  - Provide information
    - Discuss communication challenges distinctly from communication modality issues
  - Provide (credible) resources-
    - Firstsigns.org
    - Autism treatment network/autism speaks
    - where to go for further evaluation of concerns?

Still not sure?

-Uncertain about necessity of referral? Maybe child will “grow out of it?”

-Unsure where to refer?

-Anxiety about parent response

-Previous negative experiences referring for similar evaluations

- -waitlists
- -providers who don’t understand

-Outside of scope of practice?

Where to refer?

-Primary care physician or developmental behavioral pediatrician

- A licensed psychologist trained in ASD assessment

- University Center on Excellence in Developmental Disabilities

https://www.aucd.org/directory/directory.cfm?program=UCEDD

- Autism Society directory

https://www.autismcolorado.org/list-of-resources

Improving the diagnostic evaluation through consultation

-Provide information to diagnostic team regarding behaviors which fall outside of expectations based on child's hearing loss

-Consider sending familiar interpreter/teacher if child has idiosyncratic communication

-Refer to center that utilizes multidisciplinary approach

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Resources

Autism Society http://www.autism-society.org/

Autism Speaks/Autism Treatment Network

www.autismspeaks.org

Community Centered Boards (CCBs)

http://www.colorado.gov/cs/Satellite/CDHS-VetDis/CBON/1251586997819

www.firstsigns.org

CDE Guidelines for educational identification of ASD

http://www.cde.state.co.us/cdesped/guidelines_autismedid

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*

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Raising and Educating Deaf Children e-bulletin July 2016

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