BEST PRACTICES FRAGILE X: SPEECH and LANGUAGE

Vicki Sudhalter, PhD            Mouse Scharfenaker, MA-CCC, SLP
Brooklyn, NY                        Developmental FX – Denver, CO

A Day of Best Practice

Today we’ve shared “Best Practices” in occupational therapy, behavior, education and cognition, and now, on to our last area, speech and language

From X in the genes to X on the treatment map!

Anyone can provide an appropriate education, behavioral support, occupational therapy and speech/language therapy services for individuals with FXS based on what we know about FX specifically.
Outline of Presentation

• Some definitions
• Speech and language characteristics prevalent in individuals with FXS
• Best practice therapies

Definitions: Language

A set of rules shared by a language community that allows the members of that community to communicate thoughts and ideas.

• Phonology
• Morphology
• Syntax
• Semantics
• Pragmatics

Definitions: Speech

*Speech* is the verbal means of communicating.

• Articulation: How speech sounds are made (e.g., children must learn how to produce the "r" sound in order to say "rabbit" instead of "wabbit").
• Voice: Use of the vocal folds and breathing to produce sound (e.g., the voice can be abused from overuse or misuse and can lead to hoarseness or loss of voice).
• Fluency: The rhythm of speech (e.g., hesitations or stuttering can affect fluency).

http://www.asha.org/
Language Competencies

• **Receptive Competency**
  – Semantics
  – Syntax
  – Pragmatics
  – Morphology
  – Phonetic awareness

• **Expressive Competency**
  – Word Retrieval
  – Syntactic Retrieval
  – Pragmatics
  – Motor coordination (much of what we call speech competency)

Extralinguistic competencies that contribute to becoming a competent user of the community’s language

• Auditory attention
• Visual attention
• Inhibitory control
• Frustration tolerance
• Arousal modulation
• Ability to control social anxiety
• Working memory
• Long term memory
• Social pragmatic competency
• Problem solving abilities
• Motor control

Are these areas of strength or needs for individuals with FXS?

Phenotypic behaviors of Fragile X Syndrome that interfere with language development

• Motor delay (expressive abilities, articulation)
• Cognitive delay (e.g., inability to acquire syntactic forms within the same time frame as peers without Fragile X Syndrome)
• Sensory motor integration where the auditory environment does not cohere with the visual environment
Phenotypic behaviors of Fragile X Syndrome that interfere with language development (con’t)

• Social anxiety – where the conversational environment causes the child to become anxious
• Hyperarousal (already discussed this morning)
• Motor planning difficulty – where the child cannot form the motor planning to create the sound

Speech/Language challenges specific to Fragile X Syndrome

Speech-Language Disorders

• Most significant and pervasive characteristics of children with fragile X syndrome
• Often the first indicator to parents that something is wrong with their child
• Perseverative (repetitive) language is the most unique and defining characteristic of fragile X syndrome
Children with Fragile x demonstrate a constellation of speech and language characteristics specific to the syndrome

- Remember that FXS is a spectrum disorder and that these speech and language challenges will vary from child to child
- Understanding these specific characteristics will help you plan a strengths based approach to therapy
- Must remember the specific learning style of individuals with FXS so *diagnostically specific* intervention can be provided
- A responsible therapist is also well versed in their understanding of the effect of hyperarousal and anxiety on language

### RECEPTIVE SKILLS

- Relatively strong receptive single word vocabulary
- Reduced attention, auditory memory and sequential processing
- Poor working memory

### EXPRESSIVE SKILLS

- Sense of humor; strong verbal imitation skills
- Late developing speech
- Verbal skills positively correlated with cognitive skills
- Poor abstract reasoning skills

The majority of children do acquire some verbal skills
SPEECH CHARACTERISTICS
Fast rate
Disordered rhythm
Modulation of volume
Developmental articulation errors
Oral and verbal dyspraxia

The above speech characteristics result in a “cluttered” quality

SOCIAL LANGUAGE ABILITIES
• Difficulty with direct questions
• Phrase and topic perseveration, high use of automatic phrases
• Reduced topic maintenance, tangential comments, impulsive responses and poor eye contact
• Excessive verbalization, self-talk

ORAL MOTOR SKILLS
• low tone, decreased sensitivity, drooling
• hyper-sensitivity to textures, touch, temperature
• overstuffing mouth
• chewing clothes and fingers
• food preferences
Best Practice Interventions

Multidisciplinary Intervention

• Early intervention to maximize potential
• Programming with regular education peers, as much as possible, to provide appropriate models
• Emphasis on simultaneous vs sequential processing: whole word reading, gestalt presentation of tasks and activities

Intervention should be applicable to real life activities: the more relevant the therapy is to a child (think naturalistic, relationship based therapies using interest areas) they are more likely to develop stronger language skills

(Roberts, Chapman, & Warren, 2008)
TO MAXIMIZE GROWTH AND DEVELOPMENT, MUST UNDERSTAND THE AFFECT OF:

• Hyperarousal
• Anxiety
• Counter-intuitive learning style

............ON DEVELOPMENT

HYPERAROUSAL

Rec lang  Expr lang  Social lang  Oral motor
Comprehension  Syntax, Semantics, Speech Dyspraxia  Eye contact, Perserveration  Biting, Chewing

Stackhouse and Scharfenaker
Intervention for Speech-Language Disorders

**MUST INCLUDE:**

- Planning for motivation and attention
- Visual supports (pictures, logos, words)
- Routines
- Planning for transitions
- Adapt for learning style and proactive planning for hyperarousal

**MOTIVATION & ATTENTION**

- Multisensory input: visual, auditory, singing, touch
- Frequent breaks
- Use suggestions made by the OT to calm and focus
- Predictable routine and schedule
- Did you adapt for learning style and hyperarousal?

**ADAPT FOR LEARNING STYLE**

<table>
<thead>
<tr>
<th>Style</th>
<th>Learning Strength</th>
<th>Needs and Implications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual</td>
<td>Simultaneous learning</td>
<td>Use gestalt methods, don’t focus on sequential skills</td>
</tr>
<tr>
<td>Visual</td>
<td>Primarily Visual Learner</td>
<td>Use visual representations of tasks, skills you are trying to teach. Use a visual schedule of your activities.</td>
</tr>
<tr>
<td>Auditory</td>
<td>Excellent verbal and social imitation skills</td>
<td>Use vocal instructions and social modeling. Teach difficult concepts to a class partner.</td>
</tr>
<tr>
<td>Emotional</td>
<td>Highly emotional learners</td>
<td>Use an exaggerated affect without being overwhelming. These kids are one-trial learners. Use both highly positive and highly negative emotions.</td>
</tr>
<tr>
<td>Cognitive</td>
<td>Strong memory for routines</td>
<td>Use a visual schedule to depict routines.</td>
</tr>
<tr>
<td>Behavioral</td>
<td>Strong interest areas</td>
<td>Integrate these into developed materials to increase motivation, persistence, and stretch attention.</td>
</tr>
<tr>
<td>Social</td>
<td>Sense of humor</td>
<td>“Am I chopped liver?”</td>
</tr>
<tr>
<td>Social</td>
<td>Social drive</td>
<td>Use positive social interactions as motivators to teach concepts. Use nonverbal and relationship-based approaches to therapy.</td>
</tr>
<tr>
<td>Hyperarousal and anxiety</td>
<td>Hyperarousal and anxiety negatively affect learning</td>
<td>Work on it! Use consistent visual and auditory feedback. Use visual supports to help decrease anxiety.</td>
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</tbody>
</table>
| Executive Functions | Poor executive functioning skills | Use a schedule and represent information in manipulative or visual ways. Use visual supports to help decrease anxiety.
EARLY YEARS: INTERVENTION

• First words are sometimes as late as 3-4 years
• Encourage early motor and verbal imitation, combined OT and speech
• Intensive therapy
• Oral exploratory play and stimulation - these affect development of sound production skills
• Utilize a total communication approach - signs will fade out as verbalizations increase

In General take Advantage of:

• Strong verbal imitation skills
• Love for dramatic
• Response to rhythmic input
• Power words and phrases
• Teach parents several language stimulation techniques i.e. modeling, expansion, naming
• Hanen course (www.hanen.org)
Low and high tech devices:
Encourage language development—do not prevent verbal speech

Total communication

PERSEVERATION

• Look at the nature of the perseveration
• Monitor and anticipate anxiety level
• Redirect the child verbally, acknowledge the perseveration
• Motor response to “break” perseveration
• Limits on perseveration

EYE CONTACT

• Decreased eye contact often secondary to social anxiety
• It will improve with familiarity and comfort level
• Do not demand or train
DIFFICULTIES WITH DIRECT QUESTIONS

• Receptive language often stronger than expressive
• Direct questions and anxiety
• Comments or fill-in-the-blanks
  — “I want to go to the park...hmmmm. I wonder what I need to bring.”
  — “Ethan, over the weekend you went to visit...”
  — “Boy, you look hungry, I bet you’d like to eat some...”

SOCIAL LANGUAGE ABILITIES

• Difficulty with direct questions
• Phrase and topic perseveration, high use of automatic phrases
• Reduced topic maintenance, tangential comments, impulsive responses and poor eye contact
• Excessive verbalization, self-talk

• Many of these challenges are secondary to hyperarousal
• Use of video modelling with an important peer or relative as model to teach a variety of social language functions
Fast Rate

• Utilize proactive OT recommendations to calm & decrease anxiety
• Use whole body movements to emphasize slower rate and prosody
• Use a rhythmic voice to model rate and prosody

Rate/rhythm (cont’d)

• Language master, tape recorder, apps for slowed speech for modeling and feedback
• Work on increased self monitoring (video modelling)
• Eventually adapt a cueing system such as head nod, tap on the shoulder or sign for “slow” or “sing”

Rate/rhythm (cont’d)

• when anxious in class due to “demand” speech, utilize cloze technique
• prepare child ahead of time with questions
• allow him to use a tape recorder if needed
DYSPRAXIA

- In severe cases, utilize augmentative or alternative means of communication
  - LAMP Words for Life
  - Proloquo2Go
- Fully assess oral motor status including coordination of movement of the tongue, lips and jaw

Dyspraxia (cont’d)

- Assess sounds that can be produced spontaneously-use these consonants and vowel sounds to encourage motorically easy to produce words that are motivating for the individual

Dyspraxia (cont’d)

- Oral exploratory play aids in tactile localization and discrimination- increases awareness for speech postures
- Utilize explicit verbal labels and gestures to help describe targeted movements
- Mirror work
- Beware of apraxia programs that require significant touch around oral area
ORAL MOTOR ISSUES

• many and varied
• child’s behavior will “tell” you what he needs
• when eliminating less desirable oral behaviors-substitute with a more appropriate, adaptive behavior

Oral motor issues (cont’d)

• chewing of clothes, ties on sweatshirts, hand biting- reaction to increased arousal state
• chewing, sucking, biting developmentally used at all stages through adulthood to change “state”
• Low oral tone
• Sensitivity to touch

INTERVENTION

• anticipate situations which change arousal; provide necessary SI input
• use sucking, blowing, biting activities which neurologically work to calm and focus the child
B.O.B.
Biting Options Box

ORAL SENSITIVITY

Food preferences and aversions relate to increased sensitivity to certain textures, temperatures, smells and tastes

- do not intrude on child’s tactile space
- learn effective brushing and joint compression to desensitize the entire body before working in the oral area
Oral Sensitivity (cont’d)

Activities include:

– oral exploratory play
– deep pressure input through activities recommended by the OT or SLP
– foods and snacks that provide resistance and a good workout

(Children often put up less of a fight when Dad brushes their teeth due to the increased deep pressure he provides them with when they’re in a headlock)

Drooling

Often seen with low oral tone, poor lip closure or difficulty with more complex motor tasks

– decreased tongue, jaw and lip control needs to be addressed
  • introduce blowing and sucking activities to strengthen area
  • use lips (no hands) to draw licorice, cheerios or cracker into the mouth
  • provide deep pressure input to the oral area moving from the outside to inside

Stuffing

• Mouth stuffing may occur as a result of decreased sensitivity, mixed sensitivity, or in an attempt to self calm by filling the mouth - giving the child intense proprioceptive feedback to the jaw
• the excess food in the mouth is necessary for the child to tactilly perceive when his mouth is full
Stuffing (cont’d)

- tactile feedback of the lips, tongue, cheeks, and jaw may be reduced
- development of sound production skills may be poor
  - provide oral exploratory play
  - eating experiences that provide intense feedback
  - deep pressure input

THE PHENOTYPE GUIDES THE INTERVENTION FOR BEST PRACTICE

THANK YOU

Vicki Sudhalter, PhD
Head, Clinical Psycholinguistics Laboratory
NYS Institute for Basic Research in DD
1050 Forest Hill Road
Staten Island NY 10314
vsudhalter@optonline.net
www.opwdd.ny.gov/institute-for-basic-research/home
718. 494.0600