

The impact of hyperarousal and other Fragile X phenotypic characteristics on language development and production

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Outline of Presentation

- Introduction to Language
- Language Development
- Language Deficits of people with Fragile X Syndrome
- Therapy suggestions

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

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

Language Development

Language Development

The most intensive period for language development is the first three years of life.

The beginning signs of communication occur during the first few days of life when an infant learns that a cry will bring food, comfort and companionship.

Language Development

- A newborn infant begins to recognize important sounds in his or her environment. In fact, studies suggest that a neonate will preferentially suck to his language community's sounds.
- Infants begin to sort out important from unimportant sounds.
- By six months a child knows the sounds of his language.

Language Development

As speech mechanisms develop and voice matures, an infant can begin to produce controlled sounds

- Cooing
- Canonical syllable
- Repetitive syllables
- Jargon
- Words

Language Development

- By 18 months, a child should have developed a vocabulary of approximately 20 words.
- Rapid word learning begins at about 18 months.
- By two years, a child should be combining multiple words into phrases and short sentences.
- Language progresses from two and three word productions through the acquisition of prepositional phrases, tenses, complex phrases, etc.

Phenotypic behaviors of Fragile X Syndrome that interfere with language development

Phenotypic characteristics of Fragile X Syndrome that interfere with language development

- Motor planning difficulty – where the child cannot form the motor planning scenarios or scripts to create the sound
- Cognitive delay (e.g., inability to acquire syntactic forms within the same time frame as peers without Fragile X Syndrome)
- Otitis media – frequent ear infections

Phenotypic characteristics of Fragile X Syndrome that interfere with language production

- Motor planning deficits
- Hyperarousal – discussed at length earlier
- Social anxiety – where the conversational environment causes the child to become anxious

Motor Planning Difficulty

- The first issue to arise is usually a motor planning deficit
- This presents during infancy as difficulty latching, eating foods and gagging
- An infant may experience failure to thrive as a result of these motor planning difficulties

The type of therapy that addresses oral motor planning deficits is called Oral Motor Therapy

Oral Motor Therapy

- Early signs of oral motor problems include:
 - He often gags when eating
 - She fights me when I want to brush her teeth
 - He hates it when I wash his face
 - She won't let me wipe her nose
 - She overstuffs her mouth
 - He grinds his teeth

How to determine if your child is a candidate for oral motor therapy

- He has difficulty imitating the sounds of his language on command
- She chews on her fingers until there are callouses
- She puts absolutely everything in her mouth
- She mouths clothing
- He drools all the time

Specialized therapies which address oral motor deficits

- Specialized programs, such as feeding therapy, may help infants gain control over their oral muscles
- Oral motor therapies for young children have been developed by Debra Beckman and Sara Johnson
- Additional examples of oral motor therapies for children include “Prompt”
- Be aware that speech therapists are not necessarily experts in oral motor therapy

The cognitive deficits associated with Fragile X syndrome interfere with the acquisition of receptive vocabulary

How to help the child acquire vocabulary

Many linguists have argued that the human brain will begin to organize and categorize vocabulary into meaningful units once the child has acquired around 50 nouns. Thus, it is very important to assist the child to acquire as many vocabulary words as possible.

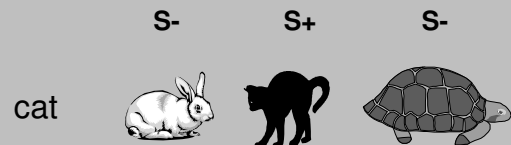
Make life harder for the child

- Parents tend to second guess their child’s needs and wants. Stop doing this. Start to make the child ask and or point to what he or she wants
- Picture Exchange Communication System (PECS)
- Fast Mapping

Fast mapping- Nouns






Baseline



Baseline




S+ S- S-

rabbit   

This panel shows a baseline trial. The word "rabbit" is on the left. Three stimuli are shown: a white rabbit (S+), a black cat silhouette (S-), and a turtle (S-).

Baseline




S- S- S+

turtle   

This panel shows a baseline trial. The word "turtle" is on the left. Three stimuli are shown: a white rabbit (S-), a black cat silhouette (S-), and a turtle (S+).

Exposure Trial #1




S- S- S+

butterfly   

This panel shows Exposure Trial #1. The word "butterfly" is on the left. Three stimuli are shown: a white rabbit (S-), a black cat silhouette (S-), and a butterfly (S+).

Exposure Trial #2




S+ S- S-

butterfly   

This panel shows Exposure Trial #2. The word "butterfly" is on the left. Three stimuli are shown: a butterfly (S+), a turtle (S-), and a black cat silhouette (S-).

Exposure #3




S- S+ S-

butterfly   

This panel shows Exposure #3. The word "butterfly" is on the left. Three stimuli are shown: a turtle (S-), a butterfly (S+), and a white rabbit (S-).

Exposure #4


S- S- S+

squirrel   

This panel shows Exposure #4. The word "squirrel" is on the left. Three stimuli are shown: a turtle (S-), a butterfly (S-), and a squirrel (S+).

Exposure #5


squirrel S+ S- S-



This panel shows a squirrel on the left, a turtle in the middle, and a rabbit on the right. Above the squirrel is the label 'S+', above the turtle is 'S-', and above the rabbit is 'S-'.

Blank Comparison or the Neither Comparison-Baseline


cat S- S+ S-



This panel shows a rabbit on the left, a black cat silhouette in the middle, and a turtle on the right. Above the rabbit is the label 'S-', above the cat is 'S+', and above the turtle is 'S-'.

Baseline


rabbit S+ S- S-



This panel shows a rabbit on the left, a black cat silhouette in the middle, and a turtle on the right. Above the rabbit is the label 'S+', above the cat is 'S-', and above the turtle is 'S-'.

Baseline

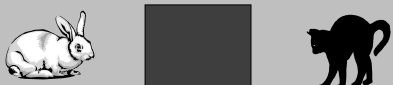
turtle S- S- S+



This panel shows a rabbit on the left, a black cat silhouette in the middle, and a turtle on the right. Above the rabbit is the label 'S-', above the cat is 'S-', and above the turtle is 'S+'.

Exposure #1


turtle S- S+ S-



This panel shows a rabbit on the left, a solid black square in the middle, and a black cat silhouette on the right. Above the rabbit is the label 'S-', above the square is 'S+', and above the cat is 'S-'.

Exposure #2




rabbit S+ S- S-



This panel shows a solid black square on the left, a black cat silhouette in the middle, and a turtle on the right. Above the square is the label 'S+', above the cat is 'S-', and above the turtle is 'S-'.




Exposure #3

S- S+ S-

cat   




Exposure #4

S- S- S+




butterfly   

Exposure #5

S- S- S+

butterfly   

Fast Mapping - Adjectives

Considerations for creating Emergent Symbolic Mapping Exposures

- Choose examples that sound very different
- Vary position of correct choice
- Vary orientation of correct choice
- Use different pictures of correct choice throughout so child does not become used to only one example

Language Production

Hyperarousal

Poor self-regulation of arousal, and the resulting hyperarousal, are important characteristics of Fragile X syndrome, and underlie many of the behavioral problems associated with it, including atypical language production.

Therapy to help a child regulate arousal

- Language is a strength for children with Fragile X Syndrome. However, because of all the demands that language puts on a child, he can find it very arousing. There are some simple actions one can take to minimize the arousal a child experiences while engaging in language.

Calming Techniques that may be used in association with Speech and Language Therapy

- Use foods with a variety of tastes and textures as well as bubbles and other motivating toys.
- Chewing on gum, aquarium tubing, etc.
- Mouth toys, harmonica, etc.
- Consider doing speech and language therapy in a group setting, which allows a child with Fragile X see models, and lets other children be first.

Consider the arousing effects of eye contact

Avoid making eye contact during conversation.

A child with Fragile X syndrome will produce his best language in situations in which he is not directly facing his conversational partner.

- While he is looking in a different direction.
- While he is sitting in the backseat of a car.
- While his parent's back is turned doing work.

Characteristic Atypical Language Productions of People with Fragile X Syndrome

Characteristic Atypical Language Productions

- Perseverative language
- Tangential language
- Repetitive and Disfluent speech

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Perseverative language

- Perseverative language is characterized by the inappropriate and/or persistent re-introduction of previously expressed and resolved topics of conversation.
- The repeated inappropriate use of favorite phrases also constitutes perseverative language.

Therapy suggestions for perseverative language

- Don't reinforce perseverative language with attention.
- Answer perseverative questions only a limited number of times.

Therapy suggestions for perseverative questions

- Help the child to de-arouse.
- Teach inhibitory control.
- Use video modeling.

Video Modeling

We recommend the social skills training videos (on DVD) produced by Terese Dana, which feature real life situations reenacted by elementary and middle school children.

More information is available about them on her web site: <http://www.tdsocialskills.com>

Tangential language

- Tangential language is characterized by utterances that do not follow directly from the preceding utterances, and the conversational partner cannot determine the associated link between them.

High Associate Combinations

Black- ----- white
Boy- ----- girl
Up- ----- down
Doctor- ----- nurse
Bird- ----- feathers
Grows on- ----- trees
Sweet as- ----- honey

Therapy suggestions for tangential language and impulsive responding

- Use Behavior Modification to teach attention and focusing
- Teach impulse control and waiting techniques (Red light/green light; Stop/Go)
- Model conversational techniques
- Video Modeling
- Self monitoring (teach the child to recognize whether he answered the question or not)

Disfluent speech

- Forms of disfluent speech include:
 - Repetition of sounds, words or phrases
 - Reformulation of thoughts within utterances
 - Self correction of thoughts within utterances
 - Placeholders (e.g., ah, um)

Examples of Repetitive Speech

- We we c c can g g go get pizza
- I I played it
- I'm I'm getting one
- Wh wh wh school do you go to?
- Whe whe where did you go to?
- What time my my my other hand?
- N n numbers
- I have I have I have watches

Therapy suggestions for word and phrase repetitions

- Teach oral motor exercises
- Teach rhythm
- Teach calming techniques
- Teach the child to slow down

In Conclusion

- Individuals with Fragile X like to communicate with others, are very empathetic and funny individuals and so language is a strength.
- We have to understand which of the phenotypic deficits of people with Fragile X Syndrome interfere with their ability to learn language and supply them with the appropriate therapy at the right time.

In Conclusion

- We have to understand how the conversational milieu engenders hyperarousal, and devise ways of making an individual more comfortable so he can produce the language he is capable of.
- We have to understand what the atypical language productions signify and provide the appropriate therapy to help that individual be more comfortable and produce the language he is capable of.