Early Communication Intervention for Young Males With Fragile X Syndrome

FPG Snapshot
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Fragile X syndrome (FXS) is the most common form of inherited mental retardation. The prevalence of FXS is estimated to be 1 in 4,000 males and 1 in 8,000 females. This study describes speech-language pathologists' impressions of the communication difficulties of young males with fragile X syndrome and the need for both syndrome-specific and individualized interventions. (Included in this *Snapshot* are a number of practical strategies.)

METHODS
Fifty-one speech-language pathologists providing intervention for males with FXS ranging in age from 2 to 9 years were interviewed.

RESULTS
Most speech-language pathologists reported that boys with FXS exhibit a visually based, experiential or holistic learning preference. They emphasized the need of making environmental accommodations for limited attention span, difficulties with topic and activity transitions, sensory deficits and low threshold for anxiety. They reported that speech goals focused on slowing rate and increasing precision for verbal children and both low and high levels of assistive technology for nonverbal or minimally verbal children. Language goals focused on listening, auditory comprehension and conversation skills. Pragmatic goals emphasized social dialogue, role-playing and maintaining a topic.

CLINICAL IMPLICATIONS
Young males with FXS present a variety of behaviors and communication impairments that are both syndrome-specific and symptom familiar. The specific communication strengths and deficits are common to many children with speech and language impairments compounded by cognitive deficits. Intervention programs should also attend closely to the specific behavioral (e.g., increased anxiety, attention deficits) and sensory "overload" problems they often exhibit before designing a tailored intervention program.

Discussion

These findings reflect a broad range of experience with a sample of boys with full mutation FXS. Findings characterize the communication issues and behaviors that speech-language pathologists may encounter, but do not reveal the actual frequency or degree of specific problems in boys with FXS in general. Practices cited here should not be seen as proof of a particular strategy nor endorsement of specific approaches. It is important to re-emphasize that though none of the children interviewed had a dual diagnosis of FXS and autism, autistic-like behaviors were often described as clinical features or concerns and this should be kept in mind as well.

In general, intervention practices emphasized recurrent themes, such as the need for visual cues and structured, stable routines. The strategies, however, represent a range of therapy techniques used across many diagnoses in which speech-language delay plays a predominant role. This range of approaches may reflect both the chronological and developmental age range represented in the sample, as well as the lack of a shared knowledge base about strategies that are likely to be useful with children with FXS regardless of age or stage of development.

How the therapists characterized their experiences with young males with FXS gives us preliminary information about learning and communication trends, as well as strengths and weaknesses in the speech and language profiles of these children. The characteristics mentioned most often include: (1) a visually based learning preference, (2) a need for structure and repetition, (3) sensory processing difficulties, (4) limited nonverbal and verbal pragmatic/social skills, and (5) erosion of speech intelligibility in connected speech.

Consideration of these characteristics when planning treatment would enhance a program tailored to a specific child's developmental level, interests, and communication priorities. Here is an expanded description of these characteristics.

VISUALLY BASED LEARNING PREFERENCE
When planning speech and language intervention activities, therapists stressed the need to provide visual cues such as photographs or "picture symbols" (i.e., simple line drawings that depict nouns, verbs, etc.) when implementing almost any speech or language goal, when trying to promote attention and comprehension, or when trying to facilitate transitions between topics or activities. Some children needed to start with more concrete visual and tactile cues such as favorite toys or miniature objects representing the next task or their turn to speak or act. Communication "books" created by the child with the therapist or with peers, teachers, siblings or parents also appeared to be very useful adjuncts to both receptive and expressive language teaching activities.

Need for structure

The children seemed to benefit from stable routines when transitioning into treatment, when moving through tasks during a session, and when ending sessions. Some seemed more disrupted by changes in the physical arrangement in the room, others by the level of light or noise, and others by changes in the sequence of events during sessions. Regardless of transitions or events that seemed most problematic, the therapists typically devised a multi-modality system to prepare the child for what would happen next. Examples included picture boards that depicted each activity and the order in which they would occur. Since the ultimate goal was for the child to transition with verbal instruction only, therapists emphasized combining verbal input with the picture cards or other visual system they were using.

Sensory processing difficulties

The recurring messages from therapists related to sensory processing reflect the need to adjust the environment so that the child would be able to complete tasks without being overwhelmed by their particular sensory vulnerabilities. Some therapists found it helpful to sit behind the child and provide gentle but consistent physical boundaries so that the child's physical impulsivity would not constantly interfere with attention to task. Other therapists found it helpful to eliminate other visual distractions or to reduce the noise to let the child complete activities. Many therapists said these accommodations could be slowly phased out for some children as they gained competence.

Limited nonverbal and verbal social skills

Many therapists put a very high priority on goals related to social routines, or on teaching simple things like greetings and polite responses, such as "please" and "thank you." Because boys with FXS often seem unaware or inattentive to the social relevance of these skills, they rarely pick such skills up spontaneously. On the other hand, when these skills are taught and words are used appropriately, such as "hi" or "please" or "thank you," at home or in the classroom, the boys are more often attended to by peers and adults and included in every day routines.

Erosion of speech intelligibility

Of the 31 therapists who worked with children who had enough functional connected speech that they felt comfortable rating their intelligibility, 28 (or 90%) rated the children as clear in single words. However, only 31% of the therapists rated those same children as clear in connected speech. Their goals for speech most often related to slowing the child's rate, emphasizing the articulation of final consonants (which has the effect of slowing rate), or practicing specific "scripts" or sentences that would be useful in the classroom on a daily basis.

Summary and implications

This is a descriptive report of current intervention practices by a relatively small sample of clinicians providing speech-language services to boys with FXS. The investigation did not use a control group nor test for outcomes based on these interventions. Therefore, the results must be interpreted with a great deal of caution.

Nevertheless, several recurrent themes were noted that may have relevance to clinical practice. For example, boys with FXS often have difficulty with transitions, either from task to task or location to location and seem to benefit from a predictable routine and structure.

They also seem to learn new concepts and communicate more successfully when visual stimuli are presented along with auditory input.

Clinicians should also anticipate weaknesses in pragmatic skills and limited awareness of nonverbal social cues. Although a minority of the boys (i.e., 15 to 20%) may be functionally nonverbal, most are verbal and tend to be difficult to understand in connected speech, despite intelligibility at the one-word level. These recurring symptoms seem strongly influenced by sensory integration issues, frequent oral-motor weakness, and behavioral vulnerabilities that also seem fairly common in this population.

In particular, clinicians should anticipate low thresholds for sensory input in one or more areas (e.g., increased sensitivity to sound, light, or touch), as well as increased anxiety in new or demanding situations. Consequently, the learning preferences, needs and strategies described here provide useful insights and starting points for clinicians when designing communication interventions for this population.

More research needs to be done to confirm the preponderance of these characteristics in males with FXS and to further delineate their relationship to developmental trajectories in these children in general and to their acquisition of communicative competence in particular. ■

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