THE FRAGILE X SYNDROME:
SPEECH AND LANGUAGE CHARACTERISTICS

By Sarah Scharfenaker, Speech/Language Pathologist

Most children affected with fragile X syndrome will have some form of speech/language delay. Because they do not speak in short phrases until 2 1/2 years of age, these children are often referred to a speech/language pathologist before a diagnosis of fragile X is made. The speech of fragile X children has been described as compulsive, narrative and often perseverative. Short bursts of speech with disruptions in the flow, including repetitions of sounds, words, and phrases are often heard. Fragile X speech has been described as "cluttered". Poor topic maintenance with frequent tangential comments may occur. Syntax is usually appropriate for mental age; a high receptive vocabulary score is usually seen, and auditory sensory and processing skills are weak. Research into the speech/language characteristics of fragile X children is ongoing and not limited to the above characteristics. Therefore, a child who has speech/language or learning problems, in addition to any other characteristics typical of fragile X syndrome, should be referred for a more formal evaluation.

Cognitive Characteristics

While many children with fragile X score in the mentally retarded ranges on standardized tests, they are often not perceived so by their parents. Many fragile X children have well-developed verbal abilities and are quick to pick up environmental facts. They are engaging and alert to what goes on around them. Females are generally higher functioning than male, though each sex typically has some learning disability. Fragile X children are usually better visual learners than auditory learners and many have auditory processing problems. These problems may be related to the significant attention deficits frequently observed. The ability to recognize and remember visual gestalts often leads to relative academic strengths in sight reading and spelling in higher functioning children, while reading comprehension is likely to lag behind. Arithmetic tends to be a relative weakness across age and IQ levels. Many fragile X children have a difficult time adapting to novel problem-solving situations and many benefit from a simultaneous, holistic approach to teaching. Because of the unevenness of their skills, they are always a puzzle to parents, therapists and teachers.

Sensory Integration Deficits

A person with fragile X syndrome often feels overwhelmed by sounds, sights, movements, touch, and even by the smells and tastes in the environment. This sensitivity to stimulation can be very upsetting, and the person may try to stay under control by constant talking (which may be out of context or inappropriate), hand flapping or hand biting, avoidance of eye contact, or withdrawal. There may be dependence on parents or other trusted caregivers for security and boundaries, both physical and emotional. A fear of movement (perhaps seen with reluctance to play on swings or slides) may be a problem as well, and the child may show a combined avoidance and need for movement by spinning toys or other objects. The problem with screening out the important from the unimportant in the environment can make it difficult to focus attention or to relate in other than a stereotypical way to another person. It may cause hyperactivity and impulsivity, tantrums, emotional outbursts, or aggressive behavior. The individual tries to cope with and make sense out of a confusing world in ways that can make his life, and yours, puzzling and frustrating.

Speech Characteristics

- Fast rate
- Disordered rhythm
- Oral and verbal dyspraxia
- Increased volume

**These speech characteristics result in a "cluttered" quality.

Language Characteristics

- Relative strengths in receptive and expressive vocabulary
- Syntactical skills commensurate with language age
- Strong verbal and behavioral imitative skills
- Strong visual skills
- Good sense of humor
- Pragmatic difficulties including problems with topic maintenance, tangential comments, impulsive responses and poor eye contact
- Phrase and topic perseveration, high use of automatic phrases
- Difficulty with semantic relation (temporal, sequential concepts, inferences)
- Poor abstract reasoning skills
- Auditory, memory, sequencing difficulties

SPEECH LANGUAGE DEFICITS AND INTERVENTION STRATEGIES

Deficit Areas	Intervention Strategies
1. Attention	Utilize child's interest area Supplement auditory with visual input including photographs Keep auditory and visual distraction at a minimum Work in small, partitioned areas Use headphones to dampen sound Calming activities provided by occupational therapist Work for short periods of time
2. Delayed onset of expressive	Utilize a total communication approach--fade out speech signs as verbalizations increase Reinforce any attempts at speech Shape responses by modeling correct utterances
3. Rate, rhythm, "cluttering"	Model correct rate/rhythm Melodic intonation therapy (Sparks et al., 1984, Sparks and Holland, 1976). Increase self-monitoring Voice synthesizer for auditory and visual feedback
4. Oral and verbal dyspraxia	Music, singing, movement Integrate therapy with occupational therapist (Windeck and Laurel, 1989)
5. Memory	Elicit attention prior to task Pair auditory information with visual cue Rhythm and music to cue recall Use short instructions intrinsic to task

6. Abstract reasoning, problem solving skills	Utilize realistic, meaningful materials of interest to the child Begin at concrete, systematically increase level of abstraction Use of microcomputers/game format Think Aloud Program (Camp and Bush, 1981)
7. Verbal perseveration tangential comments, topic maintenance	Allow increased processing time Model desired utterances Reduce complexity of utterances to child's level Monitor anxiety level and adapt accordingly (calming and focusing activities) Provide opportunities to practice a variety of speech acts Redirect the child verbally Have the child reauditorize to help process Emphasize "topic" through use of high interest materials

General Strategies

- Because imitative skills are often strong for fragile X individuals, small groups consisting of higher functioning children should be utilized to augment individual therapy.
- Maintain close coordination of goals between all those working with the child. Utilize the parents to follow through on home programs.
- Evaluate the need for combined speech/language and occupational therapies.

SPEECH AND LANGUAGE INTERVENTION

General Strategies

- Use child's interest areas in developing therapy materials.
- Use visual cues, including photographs, to supplement auditory input.
- Verbally prepare the child for changes in routine; use music to indicate transitions.
- Keep auditory and visual distractions to a minimum. Have the child sit where distractions are minimized; working independently in a small partitioned area may be beneficial.
- To help attention and focusing skills, a period of "calming" which may include deep pressure and restricted auditory input should precede and follow tasks requiring cognitive and fine motor skills. Consult the occupational therapist for specific techniques.
- Maintain coordination of goals between those working with the child. Pass a notebook between the child's teachers and therapists and have it sent home routinely for parents to follow through on suggestions at home.
- Avoid giving lengthy directions. Use short instructions and keep them intrinsic to the task. Gain the child's attention first.
- Avoid excessive talking to fill in voids in conversation.
- Allow increased processing time when the individual responds to questions or follows directions.
- Work for several short periods of time rather than half-hour or 45 minute sessions.
- Reinforce any attempts at speech. Shape responses by modeling correct utterances.
- Adjust complexity of utterances to the level of the child.
- Model desired utterances in an attempt to reduce verbal perseveration.
- Because imitative skills are often strong for fragile X individuals, small groups consisting of higher functioning children may be more beneficial than individual therapy.
- Use rhythm and music to assist in developing motor planning.
- If oral speech or total communication are not viable communication approaches, evaluate the child for use of an alternative communication system.