



Strategies for day-to-day life

A freshly brewed and delicious way to learn more about speech and language therapy and occupational therapy as used in the treatment of fragile X syndrome

The 10th International Fragile X Conference in Atlanta, Georgia this past July was a tremendous success. Why? Because the conference appeals to parents, teachers, scientists, therapists, caregivers and a whole host of other people. It unites all those in the Fragile X Community. The recent conference included 108 sessions across five days. A wealth of information was presented; it was a veritable Fragile X Frenzy!

As therapists working within the Fragile X community, we have been entrusted by parents to provide state-of-the-art services to their children. We are held accountable to understand the learning style of children with FXS, and how to best teach to that style. One of our presentations at the Atlanta conference was *Fragile X Syndrome: Strategies for Day-to-Day Life*. It outlined an assortment of strategies that are drawn from evidenced-based practice and knowledge of the neurocognitive development of children with FXS. This article is a synopsis of that presentation. We've included some handouts that we think you'll find helpful as well. *Cheers!*

Children with fragile X syndrome are often anxious and overwhelmed in a variety of situations. They may fall apart during a transition that they had no trouble dealing with the day before; they may resist a simple change in routine for no apparent reason; they may have a very successful day at school only to come home and bite or spit at Mom. These behaviors are typically *not* volitional or willful. Instead, they stem from the serious anxiety and arousal issues inherent to this disorder. Hyperarousal can lead to fear and a fight-or-flight state, causing the child to aggress, hide, run away, or refuse to comply with a request. With children in this state, we often observe diminished access to language, making it hard for them to communicate their feelings or needs of the moment.

We do find, however, that many of these challenging behaviors can be managed with a variety of strategies that are

mindfully integrated throughout the day. These strategies require a team to develop and have the most impact when followed throughout the child's day. The strategies can be broken down into three areas: Sensory-Based, Routines-Based, and Language-Based.

SENSORY-BASED STRATEGIES

One of the most fundamental sensory-based strategies is the *sensory diet*. A sensory diet is an occupational therapy intervention devised to *attain* and *maintain* appropriate arousal states throughout each day. A sensory diet consists of a carefully planned program of specific sensory-motor activities that is scheduled according to each child's individual needs (Wilbarger & Wilbarger, 2002) and each family's schedule and resources. Like a diet designed to meet an individual's nutritional needs, a sensory diet consists of specific elements designed to meet the child's sensory integration needs. It is

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based on the notion that controlled sensory input can affect one's functional abilities. Martin (1991) states in *Principles of Neuroscience*: "Sensory systems are not only our means for perceiving the external world, but are also essential to maintaining arousal, forming our body image and regulating movement."

Wilbarger & Wilbarger's (2000) comprehensive

approach to treating sensory defensiveness includes education and awareness, a sensory diet, and other techniques. One is the "Wilbarger Protocol" or "Therapressure" technique, which uses deep pressure to certain parts of the body, followed by joint compressions. These techniques employ the

proprioceptors (sensory end organs that are sensitive to stimuli) as the source of sensory input.

The Wilbargers also suggest a specific protocol for addressing oral sensory defensiveness. Either of these strategies is used in combination with an overall sensory diet, which integrates sensory motor activity into the life routine of the individual it is designed for. It is critical that this protocol not be used in isolation, and that it is initiated and monitored by an appropriately trained occupational therapist.

The *How Does Your Engine Run?* program (Williams & Shellenberger, 1994) is a step-by-step curriculum that teaches children simple changes to their daily routine (such as a brisk walk, jumping on a trampoline prior to doing their homework, listening to calming music) that will help them self-regulate or keep their engine running “just right.” Through the use of charts, worksheets, and other activities, the child is guided in improving awareness and using self-regulation strategies.

The use of this program greatly enhances the overall structure and effectiveness of the sensory diet.

We know that difficulties with sensory integration can have a profound effect on participation in everyday childhood “occupations”—play, school and family activities. Collaboration between the therapist, teacher and parents is the most efficient way to understand the child’s behavior and unique sensory needs. The “therapist-teacher-parent” team must work together to successfully implement a sensory diet and support the child’s performance across multiple environments.

Typically, a sensory diet is best designed by the family and therapist together. The therapist utilizes the direct treatment time to learn the individual child’s “formula” for attaining and maintaining appropriate sensory reactivity and arousal modulation. An example of a Sensory Diet Schedule is outlined below. *Note: The following techniques should be done only under the supervision or with the consultation of an occupational therapist.*

TIME	KEY EVENTS IN DAY	SENSORY DIET ACTIVITIES	TRANSITION STRATEGY
	Wake up	Wake up routine w/pressure input “pressure sandwich,” or joint traction while singing	
	Breakfast	Oral pressure protocol; oral supports*/heavy work	Review picture schedule of day
	Play time/gross motor time	Heavy work and play (climbing, pillow play) heavy work “chores” such as carrying items, cleaning tables (scrubbing work!), etc.	Use picture schedule
	Table top or playroom time	Wear weighted vest	
	Play time	Sensory social routine** with pressure/traction/compression input	Use picture schedule
	Lunch	Oral pressure protocol before lunch with oral supports/heavy work	
	Transition to School	Pressure play (with rhythm and music/singing if desired) and use of weighted blanket if child accepts	Transition song in car to school; weighted vest in lap
	Arrive at school	Sensory social greeting routine with pressure input	Use picture schedule
	Center time at school	Wear weighted vest	
	Play time/gross motor time	Heavy work and play (climbing, pillow play); heavy work “chores” such as carrying items, cleaning tables (scrubbing work!), etc.	
	Transition to home		Heavy work activity, then into house; use picture schedule
	Play time	Heavy work with play (climbing, pillow play)	
	Transition to quiet play time	Squeeze and hug time w/Mom or Dad; wear weighted vest during play time	
	Dinner	Oral pressure protocol before dinner with oral supports/heavy work	
	Family time	Play wrestle/pressure play time	
	Bedtime	Bedtime routine; pressure rhythm and weighted blanket	Use social story

continued ➔

* Oral supports include the use of things to bite, chew, suck, blow or get other oral sensory input from.

** Sensory social routines are play routines that simultaneously provide sensory and social input. For example, playing “This little pig went to market” with emphasis on the social sharing/anticipation and the proprioceptive input to each finger would be a sensory social routine. We often invent games, such as up-and-down play or pulling on arms and popping off the end of the hand with a fun POP! social exchange.

FIVE FINGER APPROACH

We can teach children and adults with FXS to recognize their hyperarousal and anxiety and to counteract them with the “Five Finger Approach.” This approach provides a portable and highly visual cue that we always have with us. We can assign a purpose to each of our five fingers and use the first two, or all five, depending on what fits for each child. The Five Finger Approach can either be modeled by the adult on his or her own hand or demonstrated on the child’s own hand, or modeling and demonstration can be combined.

When the child becomes upset, overwhelmed or needs to settle down, you:

- Grab your *thumb* with your other hand. This is a signal to *Stop!* (You can also verbalize “Stop!” or say, “I need to settle.”)
- Next, grab your *index finger*. This is a signal to take a “*Deep breath.*” (You can say, “Deep breath!” or “Blow hard!”)
- Grab your *middle finger* and ask, “*What do I need to do?*” (This is the time to identify that they are out of sorts and need to settle—“I am mad and I need to relax,” or “I am mad and need to tell you.”)
- Grab your *ring finger* and ask, “*How Do I Do It?*” (What strategy needs to be used? What kind of oral motor, movement, touch or visual input is preferred by the child to move into a more optimal state? You must be aware of this ahead of time.)
- Grab your *pinky* and say, “*Do It!*” Then, “do” whatever strategy was selected at the ring finger step.

While the Five Finger Approach requires self-awareness, there are many other strategies that can be implemented.

Remember, of course, that these must be done under the supervision of an occupational therapist.

READY/NOT READY CUEING

We also like to have a way for children to communicate if they are “ready” or “not ready” for the next activity, event or interaction (demand). This helps them learn to identify their “state” of preparation or readiness for the next demands or

challenges. The nice thing about this strategy is that it takes the focus off the children “not being ready” for a new activity and helps them learn how to get themselves ready (prepared) for the next task. The children are cued using visuals and modeling. The goal is to help them learn to indicate when they’re *not* ready, which importantly signals a need for them to *get* ready. Therein lies the seed of their self-regulation!

Ready-not ready cueing helps children move toward larger adaptation and coping skills. A final important point: “Not ready” doesn’t mean, “I don’t have to do this.” Rather, it means, “I have to get ready to do this.”

We use a visual support to mark “ready” or “not ready.”



It is also important to provide sensory-based coping strategies and a clear model for use of these techniques as a means for the child to move from a “not ready” to a “ready” state. In learning what “ready” and “not ready” mean, the child should have lots of exposure to the visuals and concepts. Make sure you catch your child meeting various daily demands and then indicate that he or she was “ready!” This way, they will really learn what “ready” means. Also, use the visuals yourself and when you find yourself not ready for something, label it, talk about it and teach your child through your own experience. Always pair “not ready” with clear examples of coping and preparing for getting ready to meet the demands of the situation.

ROUTINE-BASED STRATEGIES

Another avenue for keeping the day organized is routine-based strategies, which help keep anxiety about the uncertain and other arousal sources at bay. We know that children with FXS may become:

- disorganized during transitions;
- upset when new activities are presented;
- distressed when activities are altered;
- upset when an activity does not have a defined beginning and end.

Visually-based schedules are integral to maintain predictability, indicate changes and specify the “beginning” and “end” of an activity or event. Visual schedules can be complex or simple, portable or stationary, used with real objects, pictures or icons. Some excellent resources for visual schedules can be found at the following websites:

- www.do2learn.com
- www.usevisualstrategies.com

The work process systems designed by TEACCH (www.teacch.com) are another practical-routines-based strategy geared towards making the process of “work,” be it academics, play or vocational activities, highly visual, with a defined beginning and end. Again, the predictability and visual aspects of a work process help reduce the arousal and anxiety that come with the child wondering “How much work do I have to do?...How will I know when I’m done?...What do I do next?” Work processes can be comprised of a series of baskets, a vertical stack of drawers or any assortment of containers that help to visually designate the answers to those questions.

Language and social supports round out the day-to-day strategies that we use to minimize the range of life stressors associated with FXS. Our friend and colleague Marcia Braden, PhD, has made tremendous contributions to the world of Fragile X. Her understanding of the learning style of children with FXS and her ability to develop interventions based on that understanding are unparalleled. Marcia suggests that language-based supports be modified to capitalize on the learning strengths of individuals with FXS. Specifically, her description of “incidental learning” is a feature we have found can be capitalized on for day-to-day support. This learning style is characterized by children appearing uninvolved in a learning situation and physically placing themselves at a distance from the activity. But in reality, they are absorbing the information being presented. We can take advantage of this learning style when trying to shape or change behaviors through specific language and social-based strategies, including side dialogues, social stories and video modeling.

For example, when the target child is in the vicinity, two adults can have a “social story” type of conversation, what we call a “side dialogue,” to identify a situation or problem, and define options for behavior in that activity. A 14-year-old we saw often spit at clinicians when he found work too challenging. During a snack break, while the youth was playing with legos, two adults had a conversation similar to the one that follows:

Adult 1:

Sometimes my work is hard; it makes me mad and frustrated.

Adult 2:

Sometimes MY work is hard, too. When I get mad, sometimes I spit. I don't WANT to spit, but I don't know what else to do.

Adult 1:

Hey, maybe you could say, "I'm mad, this is hard!" Or you could say, "Someone help me, this is hard!"

Adult 2:


Those are good ideas. I will try to say, "I am mad, this is hard" and not spit.

The client was listening and processing this information during the break time. Subsequently he was observed during work using some self-talk: “I will not spit, I am mad!”

Social stories, developed by Carol Gray (www.thegraycenter.org), are another language support that can be used to increase predictability, thus decreasing anxiety and possible hyperarousal. When we join Marcia to see new children at consultation clinics, we send off a small modified social story prior to the first meeting as a way of decreasing the child’s anxiety about meeting new people. We usually add pictures of ourselves as well. The story we send follows:

“Hi _____. Marcia, Tracy and Mouse are VERY excited. They are going to see you in just _____ days! When we see you, we will play, look at books and have a snack. See you soon!”

Video modeling is a bit like social stories on film! Through this technique, positive social and language models can be presented by important peers or adults in the child’s environment. The video can be observed, the behaviors imitated in role play, and successful sessions can be videotaped, gradually moving from role play to more realistic environments.

Our long experience with hundreds of families with FXS has led us to strongly suggest that all three types of strategies—sensory-based, routine-based, and language/social-based—be integrated into daily life at home, school, and in the community. These strategies maximize coping and predictability while minimizing some of the common triggers for hyperarousal that, when not proactively managed, often result in poor behavioral adaptation. 

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