

Braden on Behavior

When A Disappearing Act Is A Good Thing

About a month ago a parent of two children with FXS, Arlene Cohen, suggested that I write about how individuals with FXS remove themselves from stimulating situations in order to allow for time alone and the self-preservation that flows from it.

Coincidentally, shortly after I received Arlene's request, I attended a family wedding where I had three days to observe the behavior of a family member named Tim who has FXS. As you can imagine, all of the festivities made things rather hectic. Even though Tim enjoyed most of the wedding weekend, we frequently found ourselves asking, "Where's Tim?" After looking around, we would find him in a bedroom watching *Wheel of Fortune* or an old sitcom, happy as a clam. What was behind his retreat? His need for a break from the noise and chaos – happy though it was – in order to regroup and calm himself. Invariably, he would emerge comforted and relaxed.

This is a common occurrence. The fact that people with FXS can self advocate in this way is compelling. The process of recognizing when one's own sensory system can no longer tolerate a certain level of input is called regulation. It is a skill that we actively teach younger children with FXS. The process of teaching the child how to recognize the signs of hyperarousal can often take a long time. Tracy Stackhouse, OTR, often designs programs to teach regulation. Most of the work comes from teaching the child to recognize when he or she is overwhelmed and then apply strategies to deal with those needs.

It is not at all uncommon for these individuals to go through a "Now you see me now you don't" "staging. The person with FXS may be the life of the party, expending a great deal of energy to be so, and then suddenly disappears without bravado or attention drawn to himself. The disappearance might last three minutes or half an hour - it is as if the individual knows exactly how long he needs to get centered before returning. When this process is prematurely interrupted, the outcome can be very negative.

Sometimes, when the person with FXS removes himself from an activity that is arousing and in many ways exciting, the caregiver feels obliged to engage him in order to ensure his participation. In these instances, the caregiver's need to include the person with FXS trumps the preference that the person is communicating - to simply be alone for a while. As this interchange becomes more direct and confrontational, the person with FXS becomes increasingly resolute and oppositional, and a negative outcome often results.

When the person with FXS is younger and less capable of self regulation, loud noises and busy environments often cause behavioral excesses, aggression and non compliance. The function of the behavior is to show protest and discomfort. The child soon learns that when he demonstrates aberrant behaviors that simply cannot be ignored, the adults remove him from the situation. The result is the same as is the practice of self-removal, but the means to achieving that end is less optimal. When the person is able to remove himself independently, of his own accord and on his own terms, the result is much more rewarding for all parties.

As has been discussed in prior columns, behavior can often be misunderstood and punished because the parent or caregiver does not recognize its function. Looking beyond the behavior to what it is attempting to communicate and achieve is paramount to successful programming.

The decision of a person to remove him- or herself from a difficult situation in order to avoid a negative outcome is remarkably self actualizing. In the interest of self-preservation, the child with FXS is able to avoid an embarrassing experience and thus gain a healthy measure of independence. It is a skill, like all skills, that benefits from the continued practice that is encouraged by the significant adults in a child's life.

The next time your child attempts to avoid an over stimulating activity, remember that he just may be attempting to regulate his reaction before he becomes so hyperaroused he cannot properly control his behavior. There will be time to desensitize this reaction while providing appropriate supports in the future. Allowing him to express protest in a more adaptive way can become a measure of independence and a foundation on which other skills can be built.

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Anticipate, Plan, Prepare: Behavioral Supports for FXS

During several recent school consultations it became apparent to me that we still struggle to provide a learning environment for students with fragile X syndrome (FXS) that supports their best behavioral outcomes. A number of issues continue to hinder our quest to improve students' classroom behavior.

In most cases, the focus on behavior in the classroom relates to aggression toward an object or another person. Aggression in this case is defined as pinching, grabbing, pushing, hitting and kicking. Because these behaviors are aberrant, they are more frequently identified, counted and analyzed than other more benign behaviors. While it is easy to target behaviors that are dangerous and interfere with learning, they are difficult to treat because the cause is less obvious.

One of the most frequent mistakes is to prematurely eliminate supports when the student demonstrates, in isolation or under controlled learning, a level of competence that seems to render supports unnecessary. During my recent observations, I compiled a list of supports that are important to consider when aggressive behavior escalates in the classroom.

Visual Schedule

Given that a simple change in schedule can be the catalyst for behavioral outbursts, establishing a predictable schedule seems obvious, but is often overlooked. Depending on the level of their affectedness, some students with FXS cannot tolerate any change, so they need a variety of supports to prepare for it. One such support is a written schedule that can provide a snapshot of the day's overall structure. A written schedule is ideal for someone with FXS because it utilizes many of the processing strengths we see in this population. Visual icons provide a conceptual gestalt of the day and an understanding of how events will unfold. Sometimes, after a student begins to habituate to a daily schedule, the visual support is eliminated because the team believes it will benefit the student's progress toward independence.

It is important to realize, however, that the day's schedule is critical scaffolding that supports a positive behavioral outcome. The schedule is a tool to reduce anxiety, created by a fear of not knowing what lies ahead. It allows the student to self-regulate break times and much-needed sensory input. We all rely on some form of to-do list, day timer or palm pilot, even though we may know our schedule. It is reassuring nevertheless to have what is expected of us written down somewhere, and to know if the expectation will change.

Academic Challenge

Given the cognitive challenges that students with FXS face, academic achievement is difficult for them. They learn best with high-interest materials. Many of these students

are included with neurotypical peers throughout the day and are exposed to age-appropriate interests and behaviors. They are great imitators and notice how other students behave. The literature is full of anecdotal examples of learning styles and successful learning strategies gained from this exposure.

The student with FXS may feel embarrassed and engage in disruptive behaviors when a task is presented that is uninteresting or below the norm. For example, if the task for neurotypical peers is writing about the science lab and the student with FXS is given a puzzle of Winnie the Pooh, the dramatic difference in the caliber of the task can create conflict in the student with FXS. Unfortunately, this is more typical than one would imagine.

On the other hand, if the task is too difficult and the student is simply placed with his neurotypical peers to “absorb without support,” it may be equally as embarrassing. The student may exhibit behavior that is a reaction to feelings of failure and embarrassment.

Sometimes even with support the content is so difficult that the student cannot understand the information and is frustrated that the expectation of using the support does not pay off. For example, having the social science textbook read to the student using “Kurzweil” (computer software that reads the student’s text) may not hold his or her interest because the content is too difficult to understand without adaptations.

Social Inclusion

There are a number of ways to include students with FXS in a social environment with their neurotypical peers as well as school staff. Including the student with FXS in school-related activities such as drama, music and sports provides venues where social skills can be practiced. Contact with neurotypical peers provides good opportunities to foster friendships outside an academic environment. Encouraging tasks that can be accomplished within the school community provides a sense of belonging and reinforces that wonderful trait of cooperation. Having a job with specific responsibilities can elevate self-esteem and confidence. Many jobs such as library aide, cafeteria worker, memo delivery, attendance courier, or teacher aide have proven to be successful and rewarding to students with FXS.

Scheduled Breaks

Interspersing tasks throughout the school day provides opportunities to vary sensory input. Many students with FXS require prescribed sensory breaks during the day. These breaks can be as vital as any required course. Planning sensory breaks can be challenging, so it is critical to include an occupational therapist with specific sensory integration training in developing a schedule that provides appropriate intervention. Waiting until students with FXS are overwhelmed by a task before providing a break becomes a reactive measure and usually fails because the behavior has already begun to escalate.

The student with FXS may also experience low muscle tone, which makes it difficult to stand or sit for long periods. Opportunities within the schedule to increase physical endurance can be a proactive strategy to remedy discomfort created by periods of physical activity later in the day.

Responding to a student who refuses to walk, sit or stand can be challenging. What often follows is a mix of lying down, dragging feet or falling onto the floor. This can be difficult to remedy because the student may be bigger than the instructor. It also poses ethical dilemmas regarding physical management. Establishing a proactive strategy is a more effective way to reduce the likelihood of physically managing the behavior.

Token Systems

The student with FXS often needs reassurance or encouragement to meet the demands of increased length or complexity of tasks. The use of a token board or some type of token system allows the student to understand expectations in a very concrete way. The token system is flexible and can be used to meet individual differences based on concentration and overall ability.

Often the length of a task becomes so overwhelming that the student engages in behavior that promotes escape or task avoidance. Anxiety contributes to the behavioral cycle, and often results in behavior escalation because the task length and expectation has not been communicated to the student. The token board is a proactive measure that provides necessary support to reduce anxiety and communicate task expectation in a systematic and non-threatening way.

Another benefit of the token system is it provides an opportunity to delay reinforcement. Typically when a new task is being taught, reinforcement is delivered immediately, sometimes even if the student attempts a response. Later, after the task becomes more complex or lengthy, it is not always appropriate to reinforce every trial or portion of the task. Building a work ethic and stamina requires less frequent reinforcement, which in turn increases the expectation before reinforcement is delivered.

The token system can be used at school, in the community and during work tasks of any nature. It is an easy strategy to implement and can be portable and available in any venue. Building task tolerance, length of engagement and work stamina are all skills that can be acquired using this system.

Transitional Object or Activity

I discussed this strategy in the winter, 2005 issue of the *Foundation Quarterly*. ("Have Purpose Will Transition," <http://www.fragilex.org/html/news2005.htm#>). It is an integral behavioral support that needs to be included here. The idea of numerous transitions during the day can be daunting to a student with FXS. The anxiety created by the idea of shifting focus, location and task in a matter of minutes can be debilitating.

When one thinks about a transition, it raises obvious questions. Where will we go? What is expected? How long will I be gone? Will there be a new teacher there? How crowded will it be? These questions can all heighten concern and apprehension, which are difficult to diffuse or redirect, especially during the transition.

A proactive strategy can help the habituation process. Using something as simple as a pass or card picked up prior to the transition can set the stage for the change and redirect focus. An activity such as carrying a crate of newspapers, stack of books or folder of memos can also shift the focus from the transition to completing the task, which brings the student into the next environment.

Providing supports throughout the school day is beneficial because it allows fewer opportunities to exhibit behaviors such as protest, frustration, embarrassment and confusion. These remedies can be easily integrated into the day and take less time to implement than a succession of behavioral interventions.

The strategies discussed here are by no means all-inclusive and should be used as a template upon which others can be added. The goal is to be proactive in effectively supporting the student and reducing the need for behavioral interventions.

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The Many Forms of Avoidance

I have written a number of articles about the function of avoidance and its connection with anxiety. This has been a topic in many behavioral articles because it is so prevalent in the population with FXS. Often, avoidance is linked with aggressive behavior, especially in those who are more impacted by the FX gene and those who carry the burden of significant anxiety. The fact that they are unable to process and successfully reduce their anxiety causes them to respond to it in an aggressive manner just to avoid or quickly “get over” the feeling.

This article examines the issue of avoidance and posits a variety of ways it manifests in those with FXS, through a wide spectrum of affectedness.

Avoidance does in fact serve the purpose of escape—or at least of prolonging the inevitable. Usually the individual with FXS avoids something that is novel and unfamiliar or uncomfortable. Not having prior experience with a specific situation, person or activity becomes so aversive to them that it is difficult for parents or caregivers to coax their cooperation.

The level of a person’s affectedness tends to dictate the sophistication level of the response. For example, a male with significant affectedness may hit someone to avoid a specific event. A less affected female who may be afraid to initiate a telephone call, rather than simply refusing to call, will do so at a time when she knows the other party is away from home so that she can leave a message and not have to talk directly to the person. It may be inevitable that she will eventually receive a call back, but that is not her immediate concern. The motivation is to avoid the initial contact. In either case, the situation can leave much lament and even shame in its wake.

Avoidance behavior can range from physical aggression to shyness, exaggerating, excuse-making, lying, or physical absence. Understanding how these behaviors are linked to avoidance has been extensively studied in those who are more significantly affected because the behaviors are more overt and dangerous than other more subtle avoidance behaviors. It is, however, just as important to understand how less overt behaviors become coping mechanisms for those less affected by the Fragile X gene. Providing such understanding and tying it to more adaptive strategies is essential to these individuals’ ability to function and gain access to a less restrictive community.

Often, avoidance behaviors can be so subtle that their significance is lost. There are times that the excuse-making can become so habituated that those hearing the excuses lose perspective and regard the person making them as dishonest and untrustworthy. Avoidance can thus spiral in others’ eyes from a mere manifestation of anxiety into a character flaw innate to the subject’s personality. This is tragic for those who have simply engaged in these behaviors as a coping mechanism to mitigate their fear and anxiety. Because the behavior is subtle and

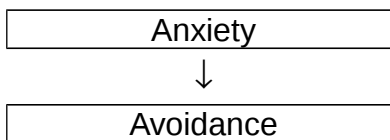
complex, it can be misapprehended, preventing it from being targeted as a behavior requiring strategic intervention.

Many individuals with FXS try to avoid disappointing others around them. This creates such intense feelings that their need to avoid overrides any concern about the consequences that may follow. For example, a person might disregard rules that lying and exaggeration are wrong because the need to please someone is stronger. The intensity of the feelings associated with disappointing others drives the behavior.

It is not unusual for me to be consulted about individuals with FXS who have made purchases for friends using family credit cards, or who have taken things from the family home in order to keep a friend. Myriad outcomes can result from this behavior.

Although it is important to remediate aggressive behavior, the more subtle attempts to avoid unpleasant matters sometimes have more far-reaching consequences. It is not uncommon for females with FXS to become so anxious that they are unable to leave their homes, participate in social events, drive, attend school or even eat or sleep. The end result can be just as harmful as a physical outburst, and it ultimately must be resolved.

Behavioral Manifestation of Anxiety



Overt	Subtle/Less Overt
<ul style="list-style-type: none">• Hitting others• Self-abuse• Running• Throwing things• Destroying property• Cursing/yelling, etc.• Crying• Refusals	<ul style="list-style-type: none">• Lying• Making excuses• Exaggerating• Hiding• Staying home (agoraphobic)• Somatisizing• Withdrawal

An essential difference between these two types of responses is that overt behaviors require assistance from the environment or another person to provide the reprieve from the situation that is creating the anxiety. The remedy is simple

because it is based on teaching the individual with FXS how to show and talk about the anxiety. Providing alternative replacement behaviors is the last step in the process of remediation. Once the person with FXS accesses more acceptable ways to deal with anxiety, it is easy to teach replacement behaviors that will garner a more positive outcome.

The less overt responses can be much more difficult to modify because they are more closely aligned with socially appropriate behavior and harder to target with immediate certainty. For example, a person hiding from unpleasant or uncomfortable experiences is much easier for others to ignore than is another person who is biting his own arm. Overt and unusual behaviors always garner the lion's share of attention.

It is also more difficult to link the behavior with the avoidance function in the case of the less overt behaviors. In addition, the person with FXS has often learned how to incorporate the behaviors into his or her personal repertoire, which tends to be accepted as a character flaw, which in turn allows for more slack to be cut. Such flaws can actually be perceived as charming—behavioral quirks that all people exhibit to one degree or other.

It can also be more difficult for the individual to accept remedial therapy for subtle avoidance behavior, because it is perceived to be intrusive. The person may have a higher level of skill development and therefore will use more subtle ways to fight the intervention, replacing one avoidance behavior with another until the pressure to change is removed.

The problem of avoidance in those with FXS will not be solved in this article, but it is important to paint the avoidance behaviors with a broader brush, including those that are less overt. The need to understand the function of these behaviors—whether overt or subtle—is critical in the management and remediation of avoidance.

Anxiety Workshop for Advocates...What are the Triggers and What are the Remedies?

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Thanks to some hard work behind the scenes by NFXF staff prior to the 2014 conference in LA, we were fortunate to have the opportunity to work with a large group of wonderful people affected by fragile X syndrome (FXS). The aim of our workshop was to offer older adolescents and young adults the opportunity to participate in the conference for the first time at these meetings in a stream specially designed for them. Most have struggled with educational settings in the past, so it was truly an amazing experience to see this group of participate with such high levels of concentration and for extended periods of time. They gave thoughtful presentations, asked appropriate questions and responded with great empathy and humor.

Over 40 male and female advocates with fragile X syndrome (FXS) from age 17 and older participated in this workshop which discussed anxiety and how to manage it. Some gave formal presentations about their own struggles and enlightened us about triggers and responses. When people experience anxiety or stress, they often experience physical symptoms which may include many of the symptoms described by the advocates. During times of stress, transition, or frustration the sympathetic nervous system response is activated by the brain, and the body responds by undergoing physical change to meet the "Fear, Fight or Flight" demand. The neurobiology of FXS includes an increase in connections within a part of the brain called the Amygdala which is part of the Limbic system involved in emotional responses. This means that most advocates are at much greater risk for hyperarousal and experience an increase in symptoms of anxiety.

At the start of the workshop, the advocates were given color coded slips of paper divided in to three categories:

- 1) Antecedents/Triggers (Pink)
- 2) Physical Symptoms (Green)
- 3) Behaviors (Yellow)

The pre-typed options allowed those who were less verbal or less able to write, the ability to choose which best applied to them. As each advocate chose those features that best described their struggle with anxiety, we opened the discussion. The following lists are in order of frequency and provided the catalyst for further discussion.

1) Antecedents/Triggers

- Being rushed
- Unexpected change
- Public speaking
- Meeting people/making social contacts
- New routine, plans
- Making decisions
- Noisy or crying kids
- Loud arguments
- Loud noises
- Phone calls
- Starting chores or jobs
- Driving
- Flying
- Navigating and finding places
- Leaving the house
- Failing
- Making a mistake

2) Physical Symptoms

- Sweating
- Fast heart rate
- Sleeping more/less
- Headaches
- Stomach problems
- Flushed faces and ears
- Gagging, trouble swallowing
- Itching
- Muscle cramps
- Trouble breathing
- Tightening of chest, feeling pressure

3) Accompanying Behaviors

- Avoidance
- Cranky/irritable
- Texting or fiddling with phone
- Obsessions, repeating a phrase
- Aggression/self-abuse
- Hair twirling
- Lying

- Biting nails, scratching, rubbing ears, picking skin, scabs etc.
- Changing subject
- Pacing/crying, hitting, fidgeting
- Laughing at inappropriate times
- Swearing

As the session continued, the advocates received information about the use of medication to treat anxiety symptoms. There are many medications that are very effective in managing the symptoms of anxiety. These have best effect when used together with behavior management strategies. Some key ideas about anxiety responses were reviewed, and then the advocates had an opportunity to practice relaxation exercises. In order to assist the process, a video was played where a self-advocate provided a model of using a visual flip chart to practice calming down. Then, the same flip chart book was used to assist the advocates in practicing the skills. The presenters walked around the room to facilitate the correct practice of the skills. Everyone seemed to benefit from having an opportunity to practice deep breathing techniques interspersed with some Progressive Muscle Relaxation techniques. These two techniques can be very effective in managing symptoms of stress and anxiety.

Because breathing exercises and Progressive Muscle Relaxation exercises can be very helpful for calming the body and the mind, the advocates were able to see how agitation and “melt downs” could be avoided. These strategies are most helpful when they have been practiced regularly by an individual before they are needed. It is helpful to practice the strategies when an individual is calm.

There are other visual tools that were used during the workshop to help the advocates rate the intensity of their feelings. These are referred to as “Feeling Thermometers” which can be used to rate the intensity of any feeling state (Happy, Sad, Mad, Scared, Worried). The Incredible 5-Point-Scale, (Kari Dunn Buron, 2012) was used to provide a visual model for a feeling thermometer and one of her work sheets was used to help the advocates understand how taking a “feeling temperature” could help them decide what to do.

Using the Incredible 5 Point Scale, the advocates were able to identify when they were at a 5 (high rates of worry/anxiety) and how they could “Try to keep quiet, put on the brakes, keep calm and carry on and “call the doctor.” They generated strategies to accompany the scale going from 5-1. When they were at a 4, they described it being time to “walk away, use deep belly breathing, put their head down” When they were at a 3, they might find it helpful to “listen to music, call parents/friends, stretch or pet a dog” When at a “2”, the advocates might “rest, listen to a happy song, take a ride in a car” When at a “1” the advocates might eat their favorite food, sleep, watch TV, swim or take a walk.

Following the exercises, the advocates also had an opportunity to review important friendship and relationship information provided by a genetics counselor. This section really provided the presenters with insight into the importance of bringing advocates together to hear about each other's experiences and concerns. It was clear that most of the participants have limited opportunities to meet with other advocates with FXS on a regular basis, and they really benefitted from sharing stories and insights into what works for them.

Understanding that there was a neurobiological reason for their struggles with anxiety seemed to provide them with the support they needed to press on. The camaraderie of the group met a need many had never addressed to share an experience that was driven by their common etiology. This reaction was a surprise to everyone.

In future workshops, the presenters are interested in expanding the scope of the format. The advocates asked that we explore relationships related not only to friendships but dating and fostering a long-term relationship with a partner. We hope to foster that type of discussion and provide more resources to support their requests. We are deeply grateful for the opportunity to pilot this session and thank each of the participants for sharing their stories with us.