

The Uniqueness of Females with Fragile X

Dr Marcia Braden

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Rood End House, 6 Stortford Road, Great Dunmow, Essex CM6 1DA

Tel: 01371 875100 Fax: 01371 859915

Email: info@fragilex.org.uk Website: www.fragilex.org.uk

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A Special Report

BRADEN ON BEHAVIOR Marcia Braden, PhD



The Uniqueness of Females With FXS

Virtually all of my previous columns have addressed in one way or other the behavioral issues of males with fragile X syndrome. That is largely because males experience both greater frequency and severity of symptoms. But females with FXS do present a variety of challenges as well, so this special expanded column will be dedicated to addressing some of them.

Females with FXS show a high frequency of avoidant behavior, mood disorders, attention deficits and learning disabilities. They are significantly more withdrawn and depressed than their typical cohorts. This makes them most vulnerable to social anxiety and avoidance.

One of the most debilitating symptoms associated with FXS in females is extreme shyness. It is not unusual to see these young girls hiding behind their parents, crying when asked to participate in conversation, unable to separate from parents and whispering or even appearing mute in social settings. They seem to need consistent coaxing in order to participate in daily life. Desensitization through repeated exposure may be required in order for them to tolerate group activities.

As these girls grow older, their shyness often translates into significant social anxiety. This anxiety can manifest in a number of problematic behaviors that result from feeble attempts to cope with its effects. For example, it is common for such girls to agree to a social plan and then spend the ensuing days physically ill with stomach distress or headaches because they're terrified of participating and don't know how to get out of the obligation. Parents often find themselves in a position of running interference by making excuses for their daughter or attempting to explain the illness to others. The problem persists as the girl faces further humiliation about why she continues to let others down. Many girls have told me they know when they want to decline an invitation but fear the fallout from saying no.

Solutions to this dilemma include the girl memorizing a set response when she wants to say no. This allows her time to separate from the intensity associated with the expectation. Phrases like "I'll have to check my schedule," "I need to see

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what my family plans are," or a simple "I think I may have a conflict" take the harshness out of saying no and afford the girl more time to think it through.

SELF-INJURY AND BULLIES

Anxiety also manifests in certain aberrant behaviors such as self-injury. Although not as severe as similar behavior seen in males with FXS, girls tend to self-injure in more subtle fashion. For example, what begins as picking a scab can evolve into a persistent picking at her arms or legs, causing scarring and extreme discomfort. The compulsion is so powerful that the

girl is often unable to stop. She then becomes very self-conscious of the behavior and often tries to cover her arms with long sleeves or cover the picked scabs with band-aids.

Girls with FXS also report being bullied or isolated from others in school. Being unable to read social cues or to understand social consequences fuels their alienation and avoidance. Female peers are often more socially motivated, which can make them less accepting of those with FXS. When these conflicts manifest, girls with FXS tend to lack the confidence to confront their peers or express their feelings of hurt. Because they seem to be more sensitive to criticism and have difficulty communicating their feelings, they are perceived as weak and vulnerable. This stance does not bode well for maintaining a position of power and self-sufficiency. The sad result is they can become convenient targets for bullying.

Because many of these behaviors result from neurobiology, it is often good to consult a physician. Many girls are helped significantly by SSRIs and other anti-anxiety/depression medications.

Also effective in building social assertion is providing concrete phrases for the girl to assert a position and then videotaping her responding to a specific event or vignette using the phrase she has practiced. Another tool is to watch a favorite movie and pause it to discuss the nonverbal behavior of the characters. This may provide the girl with helpful cues to assist in adjusting her own behavior.

ATTENTION, SPEECH, ACADEMIC STRUGGLES

Girls with FXS also struggle with attention deficits that further complicate their social interaction. Being distracted or forgetting pertinent information from a conversation does not foster social viability.

Perseverative language is often a hallmark of conversation in girls with FXS. As the girl becomes more emotionally scattered, she tends to repeat key phrases or may attempt to emphasize a certain aspect of the conversation over and over. The lack of emotional regulation feeds this tendency and causes the listener to become bored with the conversation while dismissing its merits.

Solutions to this problem may include videotaping conversations in a therapy session, providing the girl an opportunity to watch and hear the repetitious language. Sometimes it helps to use a counter to document the number of times certain phrases are repeated, thus making the intervention more concrete.

Academic difficulties may cause girls with FXS to lack the confidence necessary to succeed in school. Many studies report deficits in attention in a global sense and math in particular. These deficits are common and often pervasive, with a negative effect on school performance. Often, the deficits are not identified or diagnosed until later years, and remediation is thus delayed.

Difficulty with math can also affect the girl's ability to understand the use and value of money. Managing money and being able to budget often pose great challenges to girls with FXS. Parents often report that their daughter spends money on frivolous items and often tries to "buy" friends by spending money on trinkets and other gifts for peers.

Possible solutions: Teach the girl how to track money using a software program like Quicken. Limit credit card use, changing credit cards into debit cards only, and organize spending by using envelopes marked with budget items. Sometimes it is best to cash a paycheck and then divide the cash into categories in separate envelopes to pay the bills in a very concrete way.



EXECUTIVE FUNCTION AND NOVEL SITUATIONS

Lack of executive functioning (the ability to sum up intention, form a plan, and execute it) is another characteristic commonly associated with FXS in girls. Lack of follow-through becomes a concern as completing homework and keeping commitments eludes them. School staff may see the girl with FXS as lazy and unmotivated. Providing visual supports to explain the planning-and-executing process often helps promote completion.

Keeping a digital calendar on a smart phone with alarms set as reminders can be a helpful strategy for girls to remember scheduled events and appointments. Using iPhone applications such as Cozi enables family members to add information and reminders from other devices to keep schedules, grocery lists and assignments current.

Many parents report that their daughters have difficulty with novel or unfamiliar tasks or activities. Fear of risk-taking can cause them to avoid engagement in new activities. Routine is welcomed because it is predictable and reassuring. As a matter of fact, routine can become so addictive that elaborate rituals and compulsive behaviors become enshrined. Preserving sameness is soothing, and the girls tend to perpetuate doing things the same way over and over as a strategy to help them

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remain calm. Obviously, this coping method competes with the human need for variety and new experience, and thus causes its own stress.

Possible solution: Schedule a novel activity that is well supported and then follow it with a routine task or activity. This can help defuse fear and foster motivation to tolerate the novel activity.

Unfortunately, the need to preserve sameness does not manifest as a skill in personal organization. Quite the opposite, actually, as girls with FXS frequently have great difficulty keeping their rooms organized, cars clean, or personal items

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stored in an orderly manner. The obvious neurobiological culprits are attention and executive function deficits.

Common sense solution: Providing less is often more. Unfortunately, girls with FXS often love collections and saving mementos such as ticket stubs from football games and concerts. These and other paraphernalia make it more difficult to purge unnecessary clutter. Providing a designated place for articles of clothing, shoes and personal items helps keep things organized. Having a crate in the car for items that may be occasionally needed is also helpful. A daily list

of chores can be posted on a white board as a simple visual reminder of what needs to be done before leaving the house.

CAPITALIZING ON CREATIVITY

Many females affected with FXS are excellent writers who create wonderful stories. One woman explained that she was much more able to express emotion through a fictional character than through her real life experiences. Analogies and abstract literary techniques allow for expression of inspiring imagery and interesting stories. These girls are usually good readers who enjoy fantasy and fictional stories. Using a journal to write down their feelings is often a successful therapeutic tool.

Others nurture their creative tendencies through art forms, crafts and home decorating. Younger girls often enjoy paint-by-numbers, word searches and pattern drawing. It appears that the



structure is welcomed, providing them a sense of closure when the task is completed. Here is where the universality of art can hold great riches for girls with FXS. Parents and teachers do well to nurture these impulses whenever possible.

THE RELATIONSHIP CHALLENGE

Relationships pose a significant challenge throughout the life span. Teenage girls can become enamored with pop stars, musicians and actors. They often fantasize romantic relationships and at times even believe they have a secret relationship with a celebrity.

Their issues related to shyness and inability to express feelings complicates relationships. They may gravitate to males who have ulterior motives because the males are attentive and initially offer emotional support. This is likely to result in manipulation and exploitation of the girl with FXS.

Allowing girls to experience the "hard lessons" of life and apply them to relationships can be risky and may even estrange parents from their daughters. Giving advice or limiting access to a male friend only provokes feelings of resentment. (Yes, this may all sound virtually identical to typically developing teenage girls, but girls with FXS are far more vulnerable on any number of fronts, without ready access to typical defenses that can ease their passage through this difficult developmental period.)

The idea of having a boyfriend is very powerful, as it makes a girl feel both "normal" and "special." This can bring angst to families and often requires specific intervention from a person trained to be impartial. Allowing the girl to talk with a therapist gives her another voice of reason that she may feel is less judgmental.

Some parents insist that the boyfriend attend therapy with the girl in order force discussion about responsible behaviors. Parents have had to limit access to money, credit cards and



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checkbooks when they sense manipulation. These challenges—although not entirely unique to girls with FXS—can bring much distress to families.

On the other hand, some girls fear relationships, getting married and having children. They liken their future lives to the one they know well—perhaps riddled with difficulties and disappointments associated with a family member even more affected than they are.

This complex thicket of relationships has no easy solution, which reminds us again that the typical population has its own lifelong relationship challenges!

Strategies for addressing those challenges in girls with FXS may include establishing “dating rules” such as: 1) Never give out your phone number or address until the boy meets your parent(s), 2) Be friends first and then you see how you both feel about deepening the relationship, 3) Never give a boyfriend your checkbook or credit card, and 4) Split expenses when you go out.

Attentive parents can help set a tone for all this by spending time with and taking an active interest in the prospective “boyfriend.” Invite him to dinner, movies and other family activities. This helps ensure sure the friendship develops on solid and observable footing, not in isolation from broader family relationships, norms and values.

LOST IN SPACE

Girls with FXS usually have difficulty with visual-spatial relationships. This causes them to get lost and struggle with navigating school environments and communities. Driving can pose additional challenges related to parking, finding desired locations, judging space between cars, and remembering rules of the road. However, with good training and monitoring, such girls can learn to drive successfully, which gives them great freedom and access to jobs, social events and their communities. All this creates a positive loop for their building of confidence and self-esteem.

A possible solution for getting lost or forgetting directions is to teach the girl to use a GPS. This works well because the

voice on the device alerts her to the map and offers directions for finding her way. Also, the prevalence of cell phones now allows girls to call a parent when they are lost, which helps reduce anxiety on both ends of the line!

A BRIGHTENING FUTURE

Girls with FXS often show an interest in working at daycare centers, beauty salons, animal shelters or veterinarian offices. Many attend community colleges and some earn degrees from four-year colleges. Academic environments can be stressful, but the girls often power through and accomplish meaningful goals with great pride. I have seen it happen, and they are not isolated events.

The future for girls affected by FXS is bright. They bring much joy to their families and can be very helpful to family members and friends. They often become productive members of their communities and bring understanding to those who are more affected, serving as conduits to their brothers and other relatives who may have more significant needs. The more we learn about these girls, the more effective our interventions and support structures become in assisting them to live personally fulfilling lives. ■



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The author is a psychologist and special education consultant in Colorado Springs who specializes in Fragile X. She is the author of Fragile: Handle With Care, and creator of the Logo® Reading System.

Contact: info@marciabraden.com