

*1. Behavior, Mental Health and Medications***Medication, Behavior, and Fragile X Syndrome**

By Andrew Levitas, MD

The use of medication in the treatment of the behavioral problems associated with fragile X syndrome (FXS) begins with an understanding of the behavioral characteristics of the condition.

CHARACTERISTIC PATTERN OF BEHAVIORS IN FRAGILE X SYNDROME

Full Mutation	Premutation
Attention deficit/hyperactivity disorder	Attention deficit/hyperactivity disorder
Autism spectrum disorders, with following conditions:	Avoidant disorder
Panic disorder, other anxiety disorders	Major depressive disorder
Mood disorders	Increased risk of social deficits, shyness, anxiety, mood disorders
Aggression (primarily in adolescent and adult males)	

Of these disorders, attention deficit/hyperactivity disorder, anxiety disorders, and mood disorders can potentially be treated through the use of medication.

In attention deficit/hyperactivity disorder (ADHD), motor overactivity (sometimes seen as aimless activity or exploration), inability to concentrate or focus, distractibility, and inability to inhibit impulses (alone or as part of FXS), may continue into adolescence or adulthood.

Medication for these behaviors, often begun in childhood, can sometimes be successfully tapered or discontinued in mid- to late adolescence when hyperactivity/distractibility/impulsivity declines. Where this is not the case, persons with FXS can continue to benefit from these medications into adulthood.

To some extent, the behaviors typical of ADHD overlap with those seen in adolescents and adults with anxiety disorders, making it difficult to distinguish between them. The person with an anxiety disorder may have any or all of these symptoms, plus difficulty sleeping, obvious fright in some situations, or signs of emotional arousal not seen in ADHD. (For example: flushing, pallor, rapid breathing, rapid pulse, sweating, or panic episodes characterized by sudden panicky running, aggression or self-injury, accompanied by signs of emotional arousal.) It is possible for a person to have both ADHD and an anxiety disorder, evolving separately or together, making complete treatment a matter of a combined approach.

Panic disorder, with increasingly frequent panic responses to novelty, change, sudden sounds, social situations, or for no obvious reason at all, can either begin—or because of the person's adult size—become more severe in adolescence and adulthood. It is important when considering treatment to recognize the anxiety response underlying any aggression or self-injury that may be part of the panic response, and treat this by minimizing known causes of anxiety in the environment and by using anti-anxiety medications (typically the SSRIs) when necessary. Where aggression is a prominent part of the episodes, mood-stabilizing medication may also be of benefit, and can help some persons tolerate otherwise problematic side effects of SSRIs (irritability and “behavioral activation”—similar to what one feels if one has too much coffee).

Anxiety can exacerbate the effects of ADHD, and vice versa. Treatment must begin with consideration of which set of symptoms appears most in need of remediation, and which medication type is most likely to be tolerated by the patient. Stimulants and adrenergics used for ADHD, and SSRIs used to treat anxiety disorders (see below), are both usually well tolerated by adolescents and adults with FXS, and can be used together.

Obsessions and compulsions can continue from childhood and increase or decrease in intensity. When they become a problem for a person or his or her family, both behavioral and medication approaches can be of benefit. Medications are the same as for panic disorder and generalized anxiety (the SSRIs).

When mood disorders occur, they typically have their onset in adolescence or adulthood. They can take the form of depression, mania, or mixed mood states. Major depressive disorder can be recognized by sleep disturbance, appetite disturbance, loss of energy or enthusiasm for usual sources of pleasure, and either sadness or irritability. The person may take to bed, or refuse to leave home. Anxiety and obsessions/compulsions may intensify. Antidepressants (SSRIs) will also treat this disorder.

Mania, the manic component of bipolar disorder, also causes sleep disorders, usually major decrease in sleep, along with elated or irritable mood, easily changeable mood (“mood lability”), and an increase in energy and personal intrusiveness. Treatment is built around mood-stabilizing medications. Depending upon the severity of mania when it is encountered, various adjunctive medications may be necessary to treat the initial episode until the regime can be tapered to a maintenance program of mood stabilizers.

Mixed mood episodes, with features of both mania and depression, may require careful use of both antidepressant and mood-stabilizing medications. Where hallucinations or delusions occur, antipsychotic medication may be necessary. In the case of both bipolar disorder and major depressive disorder, the person may need to be maintained on mood-stabilizing or antidepressant medication to prevent recurrence.

The National Fragile X Foundation provides detailed information on different medications used in the treatment of FXS at www.FragileX.org.

Additional Medication Information

By Elizabeth M. Berry-Kravis, MD, PhD

It is important to recognize that although psychiatric diagnoses are applied to individuals with fragile X syndrome (FXS) to help describe, categorize, and guide treatment for behavioral symptoms, the symptoms are actually driven by the underlying biological disorder of FXS. Thus, individuals with FXS have overlapping symptoms in many psychiatric diagnostic categories and may not present “typically” for any one diagnosis. Further, because the underlying problem is FXS and not the psychiatric diagnosis, medications are not always effective, and when they are effective, they tend to make symptoms milder but do not fully treat the problem behavior. For instance, a person with FXS may respond to stimulants for the distractibility portion of their symptoms, but with a much less complete response than a person who has ADHD only. Thus, it is important for caregivers not to expect a complete response to medications, or to rely on medication treatment alone for management of symptoms. Rather, they should consider medication treatment as an adjunct, which can take the edge off behavior sufficiently to allow other behavioral and environmental strategies to be more effective.

Because medications do not always work as predicted in individuals with FXS, it is important to apply a systematic strategy for medication management. If a medication is not helping at all for a targeted behavior, it is not likely to work with other medications added. In order to prevent “polypharmacy” with the use of four or five medications (some of which may be of no benefit), medications that are not helpful should be weaned before replacing them with an alternative. Medication changes should be done systematically, with one change occurring at a time if possible, in order to best determine what is helping and what is not.

Many individuals with FXS have episodic symptoms. They may be adequately managed most of the time, but occasionally, when really stressed or bothered, will have dramatic outbursts. Medication increases are not necessarily needed simply because an outburst has occurred—often the outburst was situational and will not respond to medication changes until the individual is so sedated from medication as to be under-reactive to everything, including normal life events. These kinds of episodic behaviors are best managed with a combination of medication and proactive strategies designed to 1) identify when the individual with FXS is becoming overwhelmed, and to diffuse the situation before the outburst occurs, and 2) modify the environment to reduce the stimuli that are keeping the person with FXS constantly “on edge.”

It is further important to recognize that individuals with FXS tend to have many odd behaviors that are just part of the syndrome, do not represent any disorder other than FXS, and do not need to be treated with a medication if they are not getting in the way of function. Examples of this would be the tendency of individuals with FXS to talk to themselves, exhibit eye aversion, and have non-obtrusive routines. These are not psychotic behaviors, and most of the time they do not impair function and do not need or respond to medication treatment. Treatment should be focused on anxiety and OCD behaviors, distractibility, and aggressive behaviors that limit daily life.

Behavior Issues: Mood Disorders

Developed by the Project Work Group

(Note: There is a vignette on the DVD based on this material.●)

BIPOLAR DISORDER

People with fragile X syndrome can have moods that are disruptive to their daily lives. These are not just transient fluctuations in mood, or angry outbursts, but sustained changes in mood over days to weeks, with disturbances of sleep, appetite, and activity level.

Characteristics:

- Unable to sit for long
- Remains off task
- Paces aimlessly, sometimes tries to leave the building/house
- Aggressiveness
- Sleeplessness
- Irritability

Bipolar disorder can be treated, and future episodes minimized or eliminated, with mood-stabilizing medication. Blood tests are needed to check the medication blood levels, and parents and staff need to watch out for side effects. The parents should inform the work/house staff that their child is on a medication. They should have ongoing communication as mood, activity levels and sleep quality return to normal.

DEPRESSION

Depression can cause huge disruptions to the daily lives of people, but it can be diagnosed, and it can be treated.

Characteristics:

- Loss of interest in favorite activities and people
- Irritability
- Lack of appetite
- Moodiness
- Lack of motivation
- Sleeplessness
- Increased rigidity in routines
- Nervousness

Modern antidepressants can be extremely effective, but they can take weeks to work completely. Some require EKG monitoring. They also help with anxiety and compulsions, which are also common in persons with FXS.

PANIC EPISODE

People with fragile X syndrome can sometimes have a panic response to harmless but startling stimuli, or to no obvious event at all.

Characteristics

The person may:

- Become flushed
- Look frightened
- Try to flee
- Fight with anyone trying to restrain him
- React with self-injury, commonly hand-biting
- Sweat heavily
- Breathe rapidly
- Have a racing pulse

If such incidents are frequent, disruptive, or frightening, consult a psychiatrist about anxiety-reducing medication.

Behavior Support/Crisis Plan Hierarchy

By Marcia Braden, PhD

People with fragile X syndrome (FXS) are not generally aggressive. There are, of course, exceptions, and we hope that the following information helps caregivers plan appropriate interventions for those instances when aggression is a problem.

It has been our experience that aggressive behavior is usually a direct response to external factors. Clinical experience has provided clear evidence that identifying antecedents prior to aggressive behavioral episodes has significant merit in reducing aggression. In addition, there may be neurobiological factors that contribute to aggression. Therefore, it is always important to confer with a physician about medication.

Aggressive incidents are described by parents, caregivers, and clinicians as a “fight-or-flight” reaction to an environmental or social condition that persists when an accommodation is not afforded. When the condition or antecedent is successfully identified and accommodations are made, the person with FXS can begin to regulate his reaction within a more appropriate context. If the aggression escalates and eventually becomes unmanageable, it is important to follow a crisis plan that will keep the individual with FXS and other personnel safe. Such a plan is outlined below.

BEHAVIORAL INTERVENTION: A THREE-STEP APPROACH

Step One: Responding to Common Antecedents

The first step of any behavior plan is to intervene at the antecedent level as listed below, with redirection, reduced stimulation, and introduction of a distraction or an environmental accommodation.

COMMON ANTECEDENTS

Environmental

- Noisy
- Crowded
- Novel
- Transition-laden

Physiological

- Red face
 - Red ears
 - Covering eyes
 - Sweating
 - Becoming flushed/hot
 - Pacing, excessive movement, hand-flapping
-

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Social

- Introductions
- Answering/talking on phone
- Receiving compliments
- Direct questions

Step Two: Defusing Aggression

If intervening at the antecedent level is unsuccessful, it is important to institute the initial phase of a crisis management plan to defuse aggression and restore calm to the situation.

- Use fill-in strategies or side dialoguing between two staff members to set the stage.
For example: “We will move the table and then turn down the lights so we can calm down.”
- Remove any environmental obstructions that may become harmful to the client or others.
Have other clients exit the environment.
- Reduce verbal input, remain calm, and keep voice low.

Step Three: Actively Managing Aggression

If aggression continues, discontinue all verbal input, enlist staff support, and follow the active crisis plan and procedural remedy that is sanctioned by the school district or governing agency at your facility. (Note that these procedures vary in different settings, and require knowledgeable and trained staff to fully protect the well-being of the client while also shielding the facility from potential legal liability. This is one more strong argument for having well-trained staff.)

When Agitation Occurs: Using the “3 Rs”

By Jayne Dixon Weber

(Note: There is a vignette on the DVD based on this material.●)

People with fragile X syndrome can sometimes exhibit unpredictable behavior when they get agitated, including becoming aggressive. Here is a strategy—a 3-step plan and reward system, that with slight variations, can be used at home or at work.

REMOVE

First, you want your adolescent or young adult to realize that when he is feeling mad or frustrated, he should remove himself from the situation. This is not punishment—it is a way to deal with negative emotions. You may need to tell him to do it initially, but hopefully, he will come do it himself over time.

RELAX

Second, figure out activities that he finds relaxing. He could listen to music, read a magazine, or draw. Or he might just want to lie on his bed. He should have several options available in his room.

REFOCUS

Third, your adolescent should have a physical activity that allows him to refocus before rejoining the group. He may want to swing, bounce on a therapy ball, go for a bike ride, or go for a walk. Make sure he has more than one choice.

TEACHING THE PROCESS

First, come up with the words you are going to use for the behavior(s) you are targeting. For example, “Go to your room” typically has negative connotations, so try something like, “It is time to relax” or “Looks like you need to go listen to some music.” Feel free to come up with whatever phrase will work in your circumstance and that everyone can be consistent with using.

Explain the whole process and purpose of what you are doing. “Sometimes when you get mad you hit, so we are going to try something new. When I see you get upset, I am going to tell you, ‘It is time to relax.’ I am going to say it one time, and then I am not going to say anything else until you are in your room. You will go to your room for about 10 minutes, where you can listen to music, read a magazine, or do whatever else will help you calm down. Then you will go swing or ride your bike for a while, and then you can come back to what you were doing. This does not mean you are in trouble—this is a way that we have come up with for you to calm down and keep us both safe.”

The very first time you do it, walk through it with him. After you say, “It is time to relax,” both of you go to his room. Make sure he does something he likes while you engage in your own calming activity. After a few minutes, go for a bike ride, and talk about how good it feels.

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Mention how easy this was and how it will be a good thing for everyone in the family to do when they get upset. Then have him go back to whatever activity he was doing.

The next step is to practice actually going through this during a calm time, except he will do it on his own. Initially, you will tell him ahead of time that you are going to say the words, so he knows to expect it. Practice it a few times until you know he has the procedure down.

Continue to have occasional practices without telling your adolescent when you are going to begin the process. Again, practice until you think he has the routine mastered.

If you ever have incidents in the car, you will also want to practice the intervention there. The back seat is a good place to relax, but you will need to adapt this for your vehicle.

Here is one twist to this strategy: There are times when we all need to get our composure. Let your adolescent see you do what you are asking him to do sometimes. "I am feeling upset. I need to go relax. I am going to stay there for a few minutes, then I am going to rock in my rocking chair, and then when I feel better, I will make dinner." He needs to see that others can also benefit from this strategy.

THE REAL THING

After you think you have practiced enough, tell your adolescent that you are finished practicing, and that you are going to do it "for real" when you need to. The only caveat is that if several days pass and there has not been an incident, you may want to practice again, just to keep up the routine. In this case, you will want to tell him ahead of time if you plan to do that.

One day he comes home, is very agitated and raises his voice at you. You respond: "It is time to relax." Remember: do not say anything after you say this. Hopefully, he goes to his room. He may hesitate the first time, but glance or point towards his room if necessary.

After a period when you think he has calmed down (perhaps 10-15 minutes), go to his room and announce: "When you are ready, you can go on the swings or bounce on the therapy ball."

However, if he says anything negative to you, then tell him it looks like he still needs more time. Then walk away. Go back and repeat every 5-10 minutes until he has calmed down. Make sure he does his refocusing activity for 10-15 minutes. The time may vary—you be the judge. Then say, "You can rejoin us when you are ready."

REWARD SYSTEM

Provide a reward system to give your adolescent an incentive to carry on through this process. During all the practice sessions, give him something after he has refocused: a coupon or small amount of money that he can redeem for things he enjoys.

Since what you really want is good behavior all day, try this: Break the day down into three periods—morning, afternoon, and evening. Get a calendar you can devote to this and put three little boxes on each day.

If your adolescent does not have an incident during the morning period, you will put a mark in the appropriate box, and so on throughout the day. For each mark, he gets a token. So if he goes all day without an incident, he will get three tokens at day's end.

If he has an incident during one period and he goes to his room right away, he gets his token right away. If he does not go to his room uneventfully, he does not get his token for that period.

Another thing that you can do during the day is keep track of the good things your adolescent does, particularly as he relates to difficult activities such as transitions and not getting what he wants, when he wants. At the end of the day, talk to him about these things and let him know: "You can do this."

MANAGING RESISTANCE

Your adolescent's refusal to go to his room could be a problem. Depending on his level of agitation or aggression, the best idea is for you to leave the room if he will not. If you think his behavior may escalate, take yourself completely out of the picture, move to another room and say, "It is time to relax, and if you aren't going to go to your room then I will leave. When you are ready to follow the plan, you can let me know." From a distance, check back with him after a few minutes. Continue to do so until he is ready to comply with the plan.

ADAPT, MODEL, DISCUSS

It all boils down to the 3 Rs. This is a relatively simple concept to implement and is easily adaptable to most families. The difficult part is keeping the intervention from seeming like punishment to your adolescent, but keep modeling it yourself and talking about personal responsibility. If his behaviors do not stop or continue to escalate, be sure you talk to his psychiatrist or other mental health professional for more ideas.

Note: Our family worked with Partners in Behavioral Milestones, Inc. (see Resources, page 14) to develop a plan similar to this for our son Ian.

Reward Systems

Developed by the Project Work Group

Goal: For your child of any age to exhibit good behavior.

THE BASICS

Any reward system you devise has to be:

- 1) One your child understands.
- 2) Motivating (he or she has to buy into it).
- 3) One that *you* can follow on a consistent basis.

As you develop your reward system, here are some other ideas to keep in mind:

- Make sure your child knows what behavior you are expecting. It is easy to say “No” and “Stop it,” but does he or she know what “to do” instead?
- Develop a reward you think your child will be willing to work for at least in the beginning. Consider money, tokens, and doing special activities (watching a movie, getting ice cream).
- Know that most reward systems will have to change or adapt over time. Your child may get tired of receiving money (!) or tokens for good behavior.
- Some children will not have a concept for getting money/tokens and then saving to buy something. In those cases, you may want to provide an actual item as the incentive each time—such as a baseball card.
- Depending on your child, you may also want to have a “bigger” gift available when he or she achieves targeted behavior a certain number of times.
- Once you know what you want your child to do, when he is supposed to do it, and the rewards he will get for doing it, talk to him about it.
- Practice as much as you think you need to during calm periods, and reward her for successful practices.
- Give your child money or a token every time he exhibits the targeted behavior. Make sure you provide regular opportunities to spend it.
- Over time, you may be able to wean her from material rewards, with only verbal acknowledgement of her good behavior. The goal of all reward systems is to make the desired behavior self-rewarding and intrinsically motivating.
- Realize that this process may take a long time and will likely entail many frustrating moments. Be patient and creative.

For other ideas, consider an Internet or bookstore search for children’s behavior reward systems. You may find ideas for children who are “typical” that can be modified to meet your child’s needs.

Behavior at Home: A Few Simple Changes

Developed by the Project Work Group

1. Follow a routine.
2. Use a picture schedule.
3. Develop a method to inform of transitions to come.
4. Allow adequate time to make transitions.
5. Maintain a “sensory diet” throughout each and every day.
6. Participate in regular/daily physical activity.
7. Provide healthy food at regular intervals throughout the day.
8. Minimize unexpected loud noises and be aware of “background” noise in and around your home (lawnmowers, fans, traffic, trains, TV/radio, etc.).
9. Arrange opportunities to schedule your child’s preferred activities.
10. Purchase soft, cotton clothing (often thrift stores are a great source).

Many of these suggestions will require you to experiment with your young adult to see what works best for him or her. Remember, too, that just because it works one day does not guarantee it will work the next! Many variables can influence the routine of a day: illness, a sudden plumbing problem, poor weather, an unexpected phone call, an invitation to do something new and exciting. And don’t forget that some days both you and/or your child are just plain tired!

Do the best that you can, but know that most days will not be perfect. That is okay, because as your child matures, he must eventually learn to live in an imperfect world. It is your job to help him learn to do adapt to these imperfections over time.

PICTURE SCHEDULES

Picture schedules can be an important part of your young adult’s life. He will most likely need some sort of picture schedule for the rest of his life. Here are some ideas:

There are many options for the “picture.” For some, words may suffice: “Soccer @ 4 p.m.” Actual photos are another option, though it can be time-consuming and expensive to implement. “Pecs” is another option: icons that represent a word. There are several websites where you can download these icons for free (see reference section below). You can always draw your own picture. In cases where you cannot find an icon, this is a good option.

Try a desk calendar (approximately 18” x 24” size) that has room to write information for each day. Put it on your refrigerator and use whatever format your young adult prefers—words or

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pictures. You may also want to keep track of other family members' activities; use a different color for each person.

If you prefer more detail, try a three-ring binder and use heavy sheets as in a magnetic photo album. Put the times on the left and use Velcro to attach the activities on the right so they can be changed on a daily basis.

Know that a picture schedule may work one day, while it may not seem to make any difference the next. Or one day a certain type of schedule may work, and the next you may need to modify it with more detail. Have fun with this!

REFERENCES

- Beyond Autism Pecs Pictures/Icons Pages www.members.tripod.com/trainland/pecs.htm
 - www.dotolearn.com/
 - www.kidaccess.com/html/
- Or search by "pecs" on the internet.

SENSORY DIET

A sensory diet can be very important for young adults and, indeed, throughout their lives. Like a nutritional diet, it refers to a carefully planned program that helps individuals manage their sensory input so they are not put on "overload." Read up on the topic, talk to a trained occupational therapist, and see if it makes a difference. Many of the activities are also good for premutation carriers of the Fragile X gene.

REFERENCES

Raising a Sensory Smart Child: The Definitive Handbook for Helping Your Child with Sensory Integration Issues. Lindsey Biel and Nancy Peske, New York, Penguin, 2005.

Sensory Diet Activities for Home and Away: www.sensorysmarts.com/diet.html

The ERIC Clearinghouse on Disabilities and Gifted Education (ERIC EC)

The Council for Exceptional Children

1110 N. Glebe Rd. Arlington, VA 22201-5704

800-328-0272

Understanding Sensory Integration

<http://ericec.org/digests/e643.html>

The National Fragile X Foundation

www.FragileX.org

The Out-of-Sync Child Has Fun: Safe Activities for Home and School: Sensory-Motor, Appropriate, Fun, and Easy

Carol Stock Kranowitz

Penguin Group (USA), 2003

Out-of-Sync Child: Recognizing and Coping with Sensory Processing Disorder

Carol Stock Kranowitz

Penguin Group (USA), 2006

Sensational Kids

Lucy Jane Miller, PhD, OTR

G. P. Putnam & Sons, New York , 2006

Sensory Integration and the Child

25th Anniversary Issue

A. Jean Ayres, PhD

Western Psychological Services, 2005

Resources

Books—The following books are available from The National Fragile X Foundation (800-688-8765, www.FragileX.org).

Braden, Marcia L. *Fragile: Handle With Care, Understanding Fragile X Syndrome*, Avanta, 2000.

Hagerman, Randi J. MD and Paul J. Hagerman, MD, PhD, *Fragile X Syndrome: Diagnosis, Treatment, and Research*. Third Ed. Baltimore: The Johns Hopkins University Press, 2002.

Harris-Schmidt, Gail and Fast, Dale, *The Source for Fragile X Syndrome*, 2004.

Michael Tranfaglia, MD, *Medication Guide for Fragile X Syndrome*. FRAXA Research Foundation, 2000.

Websites—

Applied Behavior Analysis (ABA)

Cambridge Center for Behavioral Studies

www.behavior.org/ (there are numerous websites on this topic—talk to your doctor)

The National Fragile X Foundation

www.FragileX.org > intervention > behavioral disorders

Partners in Behavioral Milestones, Inc. (PBM, Inc.)

6412 E. 87th St.

Kansas City MO 64138

816-501-5138

www.behavioralmilestones.com/

Disability Solutions

www.disabilitysolutions.org/

Click on “Newsletters,” search for “Behavior”
