


# Fragile X Syndrome

## Etiology of Behavior Anxiety and Hyperarousal

Leslie and Lanie Kanat

Howard Center  
August 2015

Katie Couric describes Fragile X Syndrome and its connection to autism.



<http://news.yahoo.com/video/fragile-x-explained-010247619.html>


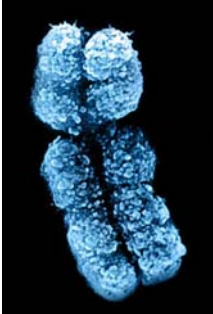
**Importance: to learn how to interact with those who have ASD associated with Fragile X Syndrome.**

**If you know why, you can figure out how.**

*– William Edward Deming  
Management Consultant*

**Fragile X Syndrome is a medical condition.**

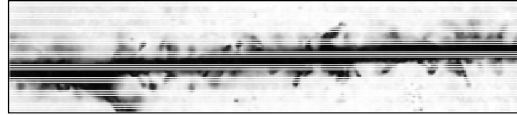
The name stems from the fact that the tip of the X chromosome looks like it might break.

<b>Fragile X</b>	<b>Typical X</b>
	

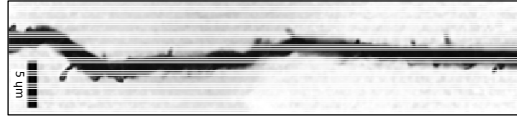
# Fragile X Syndrome is a single gene disorder.

FMR1 → ~~FMRP~~

The lack of FMRP results in long dendritic spines, increased spine density, high neurotransmitter activation, and failure of the synapse maturation and pruning processes.



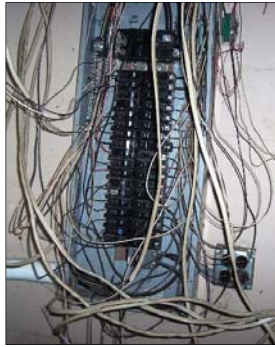
Dendrite from a human afflicted by FXS.



Dendrite from unaffected human.

<http://cercor.oxfordjournals.org/content/10/10/1038.long>

Antiquated fuse box that can not handle any more input – too many connections going everywhere.



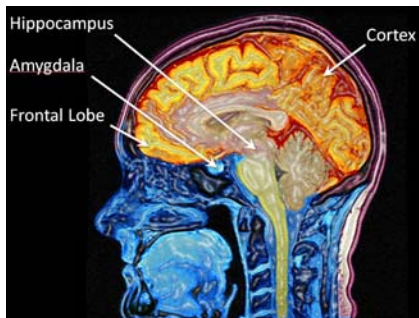
<http://www.youshouldown.com/img/homeins/panel2.jpg>

Because we can not rewire the brain, we have to make accommodations for the poor wiring.



<http://roncoelectricnj.com/files/2014/05/electricalpanel1.jpg>

The lack of FMRP results in an overabundance of improper and overactive neuronal connections.



≈60% have ASD

≈20% have seizures

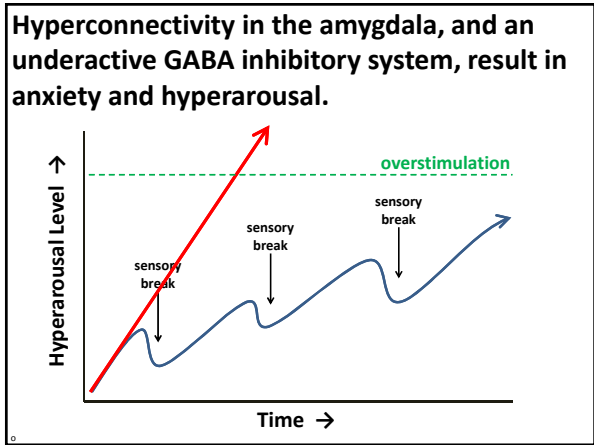
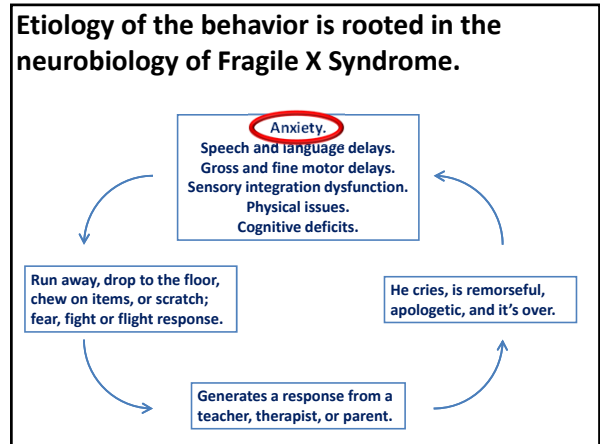
The lack of FMRP results in an overabundance of improper brain connections.

- Strengthened in the amygdala: **anxiety.**
- Weakened in the cortex and hippocampus: **short-term memory.**
- Weakened in the frontal lobe: **hyperactivity and executive function.**

**You can't battle biology.**

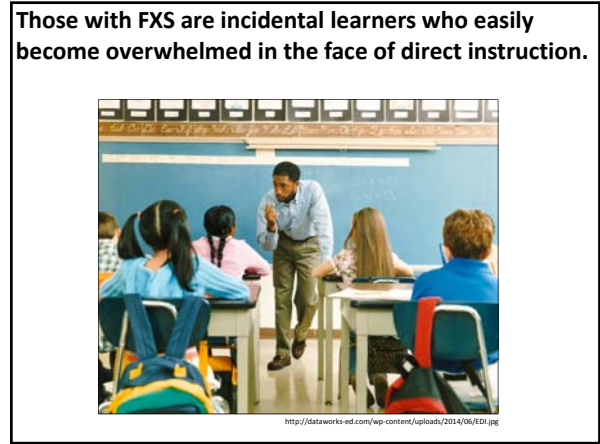
– Dr. Karen Riley

**One can't treat behaviors without accounting for the neurobiology.**



**The important issues in Fragile X Syndrome are hyperarousal and intellectual function.**

**Hyperarousal intervention strategies appear opposite in practice to the nature of many autism intervention strategies.**



Those with FXS are incidental learners who easily become overwhelmed in the face of direct instruction.

**It is critical to modify discrete timed trials, and avoid face-to-face interactions and forced responses.**

Autism intervention programs often use sequential teaching, especially forward chaining methods, yet is directly counter to the FXS learning style.



Autism intervention programs often use sequential teaching, especially forward chaining methods, yet is directly counter to the FXS learning style.

**Children with FXS are simultaneous learners. They are gestalt learners who need to see the end product. They can replicate through imitation.**

Autism intervention programs target social skills, including eye contact and social interaction, with reinforcement for correct performance.



Autism intervention programs target social skills, including eye contact and social interaction, with reinforcement for correct performance.

**When those with FXS feel social pressure, their hyperarousal skyrockets, and the social skills are sacrificed.**

Autism intervention programs target atypical behaviors (such as perseverative speech, hand flapping and biting) and attempt to modify them.



Autism intervention programs target atypical behaviors (such as perseverative speech, hand flapping and biting) and attempt to modify them.

For those with FXS it is better to proactively reduce hyperarousal rather than struggle to change behaviors.

For those with autism associated with Fragile X Syndrome, it is the FXS learning style that is first and foremost, so reducing hyperarousal should lead the way for all interventions.

ABA is highly structured, yet those with FXS need highly flexible interventions.

- Antecedent may have occurred days prior to the maladaptive behavior, or it could be a scheduled future event.
- Discrete trials do not often provide best evidence for those with FXS.
- Wait to reduce anxiety.

ABA is also highly variable in its delivery.

Dr. Marcia Braden has shown that Pivotal Response Training (PRT) and Strategies for Teaching based on Autism Research (STAR) can work with those who have FXS.

Individuals with FXS fall somewhere on the DSM-5 for autism.

- Enthusiastic
- Fun-loving
- Loving
- Gentle
- Happy
- Distractible
- Easily redirected
- Good imitators

As long as their hyperarousal levels are low.

People with autism and FXS display a **wide range** of individual characteristics.

<u>Autism Spectrum Disorders</u>	<u>Fragile X Syndrome</u>
Social indifference	Social anxiety
Gaze indifference	Gaze aversion
Appear socially passive	Generally friendly
Variable language delays	Language delays
Poor understanding of facial expressions	Understand facial expressions
Variable intellectual abilities	Cognitive delays

Thomas the Tank Engine is favored because facial expressions and directions do not change.



Etiology of the behavior is rooted in the neurobiology of Fragile X Syndrome.

- Impulsive
- Hyperactive
- Social anxiety
- Cognitive delays
- Attention deficit disorders
- Ritualistic and repetitive behaviors
- A fear, fight or flight response

Etiology of the behavior is rooted in the neurobiology of Fragile X Syndrome.

- These kids want to be normal, yet they know they are not.
- They want to please.
- Threats and coercion do not work.
- They understand failure – perceived as a threat resulting in anxiety.

Etiology of the behavior is rooted in the neurobiology of Fragile X Syndrome and therefore individuals are challenged by:

- Changes in routine
- Forced responses
- New situations
- Transitions
- Large crowds
- Social situations
- Personal boundaries

The FXS learning styles have been well documented in the literature.

- Use visual cues (not verbal cues).
- Stay neutral (and positive).
- Avoid direct eye contact.
- Provide two choices.
- Find a hook to make a connection.

These techniques will help reduce anxiety.

The FXS learning styles have been well documented in the literature.

- Uncanny social drive.
- Developed sense of humor.
- Strong memory for routine.
- Require greater organization.
- Imitate behaviors and mannerisms.
- Follow the schedule, not the clock.

The FXS learning styles have been well documented in the literature.

- Simultaneous learners (not sequential).
- Gestalt learners (show final product).
- Cloze technique (backwards chaining).
- Incidental learners (he is attending).
- Indirect instruction (involvement).
- Side dialogues (a conversation).
- Peer and video modeling.
- Self-talk.

The main problem with many autism intervention programs is that they fail to take into consideration the learning styles of individuals with FXS in which the main issue is hyperarousal.


The goal is to keep hyperarousal low.

- Use calm, simple, caveman language.
- Acknowledge appropriate behavior.
- Once you promise it, deliver it.
- There is a need for closure.
- When successful, do not ask for more, even if requested.

The goal is to keep hyperarousal low.

- Calming behaviors (self-regulation)
- Sensory diet (build in breaks)
- Speech and language therapy
- Occupational therapy
- Deep pressure activity
- Music and song

Occupational therapy helps provide proprioceptive input.



frog swing      platform swing      bolster swing, net and balance beam

<http://kanat.jsc.vsc.edu/ot/>

Occupational therapy helps provide proprioceptive input.



guitars      elliptical      trampoline

**Numerous accommodations in place at home.**

- Continuous bathroom reminders.
- Exterior doors locked from inside.
- Cabinets are locked.
- He is never left alone.
- DVD and iPad are priceless.
- Everything is planned – schedules.

**What are our goals for his future?**

- To find ways to reduce his anxiety.
- Recognize when he is becoming overstimulated and to self-regulate.
- Develop basic living skills (simple hygiene, communication, social skills, and quantitative skills).
- Find a place for him to live where he will be safe.

**Your challenge: identify those who have ASD associated with Fragile X Syndrome, and adapt the ABA approach to accommodate Fragile X learning styles.**



les.kanat@jsc.edu  
lanie.kanat@comcast.net



fragilex.org  
northernvt.fragilex.org  
fb.me/fragilexvt



fraxa.org



morgridge.du.edu/staff-members/riley-karen-s-phd



marciabraden.com



developmentalfx.org

**Live the Fragile X way.**

**Be proactive, no surprises, have accommodations in place, model appropriate behavior, and remain calm.**