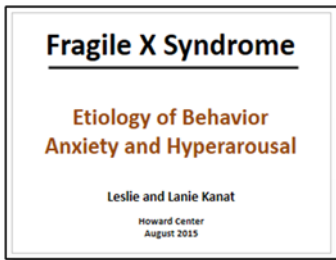


Howard Presentation Notes



We have two children: Ryan FX and Marin is neurotypical. Les has a Ph.D. in the Earth Sciences from the University of Cambridge, England, and has been a Professor of Geology at Johnson State College for the past 27 years. I have a degree in Psychology and Child Development from the University of Arizona, I was a social worker, and now I do some nutrition counseling, and take care of my family. We live in Jericho, Vermont.

Thank you for being here with us today and allowing us to explain the learning style of individuals with FXS and for being open to augmenting your intervention programs to work with those kids who have autism spectrum disorders associated with Fragile X Syndrome. We are open to any and all questions at any time during our presentation.

Many of the ideas presented today come from publications of the National Fragile X Foundation, Dr. Marcia Braden, Dr. Karen Riley, and Developmental FX whose guidance, support and ideas have been invaluable to all of us who live with FXS. Their contact information may be found in the handout at the end of the presentation.

We will start with a video of an interview by Katie Couric, with some of our friends who have a son with Fragile X, and our son's doctor from the MIND Institute in CA.

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

This phrase, autism spectrum disorders associated with Fragile X Syndrome, is the official diagnosis in the DSM-5. The DSM-5 now incorporates etiology as a specifier for neurodevelopmental disorders. Today, we will make the case for adapting ABA intervention programs for those who have autism spectrum disorders associated with Fragile X Syndrome.

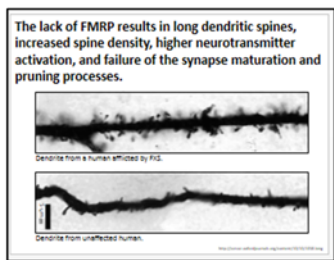
If you know why, you can figure out how.

- William Edward Deming
Management Consultant

William Deming consulted with companies all over the world to improve their productivity and efficiency. We interpret this quote to mean that we know *why* those with FXS act the way they do (it's their neurobiology), and with this knowledge we can figure out *how* to appropriately interact with them in order to improve their social skills and reduce their maladaptive behaviors.

Fragile X is a medical condition.

Fragile X syndrome (FXS) is the most common cause of *inherited* mental impairment. This impairment can range from learning disabilities to more severe cognitive or intellectual disabilities – sometimes referred to as mental retardation. FXS is the most common known cause of autism and *autistic-like* behaviors. Symptoms also include characteristic behaviors, characteristic physical features, and delays in speech and language development. It is a single gene disorder where 1 in 151 women carry the Fragile X gene.

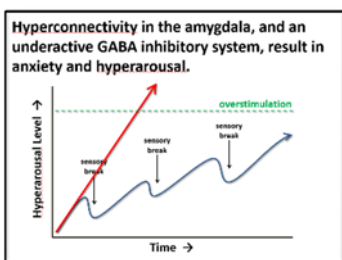


Spines provide an anatomical substrate for memory storage and synaptic transmission – they also serve to increase the number of possible contacts between neurons. Yet, there are too many connections in the Fragile X brain; there is crowding; the connections are overactive; the braking system in the brain is not working correctly; everything is

speeding too fast; too much information coming in, and this results in hyperactivity, excitability, overstimulation.

Connections in neurotypical brains become more mature, more sophisticated, more discerning, and are able to filter and prioritize information. This is not the case for FXS.

This presentation is primarily about hyperarousal, yet if we have time, and you are interested, we can talk about inheritance of the Fragile X gene and carrier status.



For those with FXS, if one crosses the threshold (overstimulation) then any kind of physical restraint will only serve to escalate a situation. The best thing to do is to wait.

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

... and this is because of the neurobiology of the Fragile X brain.

Hyperarousal intervention strategies appear opposite in practice to the nature of many autism intervention strategies. This is because anxiety plays a major role in behavior and the effectiveness of the interventions. Anxiety needs to be considered as a main factor in the design and implementation of the interventions, as well as the data collected around behaviors and interventions.

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



And it is imperative that we follow the child's lead, find their interest areas and other indications of comfort in order to increase engagement.

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



We know that children and adults with FXS make wonderful eye contact when their level of arousal is minimized, allowing them to feel comfort in the social situation. Yet when they become hyperaroused the fear, fight or flight response is activated.

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.

These kids are wired for anxiety so it is never appropriate to put these kids in situations that increase anxiety.

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

Autism Spectrum Disorders	Fragile X Syndrome
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

In this table we compare characteristics of those with ASD to those with FXS.

Social anxiety: hides from people out of place; avoid crowd

Gaze aversion: fist bump

Generally friendly: greets people from the car

Language delays: some do not have language

Facial expressions: smile gets a smile; emotive; responds

Intellectual abilities: by definition, individuals with FXS have cognitive delays, it is the Fragile Mental Retardation 1 gene that is the cause of their impairment and their symptoms.