Fragile X Syndrome Workshop
for families and professionals

Sydney, Australia
March 30, 2015
Marcia Braden, PhD
The Behavioral Presentation
Current Treatment Guidelines
Consensus of the Fragile X Clinical & Research Consortium on Clinical Practices

- Anxiety
- Social Shyness
- Hyper arousal
- ADHD
- Sensory Defensiveness
- Aggression
- Self-Abuse
- Autistic-like Characteristics
Anxiety

• This is present to some extent in the vast majority of individuals with FXS. While behaviors associated with anxiety tend to be ameliorated to a degree by standard medications, cognitively appropriate psychotherapy is an important adjunct. Modalities include but are not limited to cognitive behavior therapy, biofeedback, social thinking and the acquisition of coping skills.
Social Shyness

- Social anxiety is the anxiety created when the person with FXS is expected to respond or initiate social interaction. The symptoms can be present very early often with signs of arousal any time the young child is subjected to a social event. This can be as simple as having guests in for dinner, going into a busy environment, separating from a parent or being introduced to someone.
Hyper arousal

Hyper arousal is a state of increased psychological and physiological tension marked by such effects as agitation, anxiety, exaggeration of startle responses, insomnia, fatigue, and hypervigilance.

The key to management is to identify the antecedents that predispose towards the loss of control. These antecedents are then either avoided or, if this is not possible, calming interventions are put in place. The goal is to teach the individual to recognize the signs of hyper arousal before the loss of control, and then be able to self-calm and soothe.
ADHD

This condition is common in both males and females with FXS. Once confirmed in individuals, medication is the preferred treatment, as ADHD does not appear to be amenable to other interventions. However, it is essential to consider that other underlying issues, or simply young age, may present as ADHD. Management may therefore involve observation and/or other modalities as the assessment and management plan evolves. Evaluation usually has to include whether ADHD or anxiety is most prominent, as they often co-exist and mutually exacerbate each other. The condition deemed to be more severe is typically the one for which medication would be started first. The other condition would then be observed to determine to what degree it resolves as symptoms in the other abate. If still present, then appropriate additional medication may need to be introduced.
Sensory Defensiveness

Effective intervention typically involves a sensory program developed by an OT, focused on desensitization and/or amelioration of disturbing sensations. It is critical to include an OT who has specialized training in sensory integration disorders. The OT can design treatment based on observation as well as sensory profiles. Often, the OT works in tandem with the speech language pathologist to develop oral motor strategies as well as other activities of daily living.
Aggression

This is often a defensive behavior in FXS, and treatment depends upon the cause, which is true of any of the behaviors discussed. If aggression is severe and not responding to management strategies, then medication may be necessary.

People with FXS are not inherently aggressive and do not premeditate intention to hurt others. The aggression is usually related to a situation that has caused a fight or flight reaction.
Self-Abuse

Many individuals with fragile X syndrome engage in self-abusive behaviors, the most familiar of which is biting their hands. This results from substantial oral tendencies that cause them to put many things in their mouths (including hands and pieces of clothing). While hands-to-mouth may have begun as a form of self-soothing, the child may quickly learn that people respond to this behavior, thus changing its function to become a call for attention. One intervention method is to ignore the behavior (as long as it is not dangerous to the person). A second is to provide the individual with an alternate form of oral gratification such as a chewy toy. Eliminating the behavior requires a determination of what in the environment is creating the child’s need to self-soothe and then ameliorating the problematic stimuli.
Autistic-like Characteristics

- Autism can be comorbid with FXS. The diagnostic criteria is based on observation.
- DSM V now separates out severity into three levels which defines the level of support necessary.
- The criteria targets social language and repetitive behaviors.
How much is my child affected and what is the prognosis?
Well, none of us have a crystal ball…

- Everyone with FXS has parts of this neurobiology
- The best predictor of outcome at this point is how well the person with FXS regulated the symptoms.
Factors that Impact Behavior

- Environment
  - Cognitive Curricular
    - Low interest materials
  - Sensory Integration Disorders
  - Physical Symptoms/Anxiety
- Communication delays
What I’ve Learned About FXS and Behavior

- They show or tell us what they need
- It is our job to give them a more appropriate way to communicate their need
- We have to observe the behavior looking for function in order to help modify the behavior
They Show Us What They Need
The Behavioral Cycle

Neurobiological underpinnings
- Language delays
- Sensory dysfunction
- Cognitive delays

Anxiety

Response from Parent, Teacher, Therapist
- Running Away
- Throwing Shoes
- Hitting Head
- Pulling Hair
Foundations of Behavior

- In order to change behavior we must first understand the foundational aspects of behavior.
- All behavior serves a purpose.
- Behaviors do not continue unless they are reinforced in some way.
- Sometimes the reinforcing aspect of the behavior is internal.
- If you do not address the cause of the behavior it will not change.
Functional Assessment

• The ABC’s of Behavior
  • Antecedent (the action that occurs directly before the behavior)
  • Behavior (the specific act or behavior)
  • Consequence (what happens directly after the behavior)
ABC’s of Behavior
ABC’s of Understanding Why students engage in problem behavior: Finding out the Pay-off or Function of Behavior

A = Antecedent(s). Find out the events that occur right before the behavior.

- Allows you to predict: Where (During routine)? & When (Trigger event)?
ABC’s of Understanding Why students engage in problem behavior:

Finding out the **Pay-off or Function** of Behavior

B=Behavior. Find out what is the observable problem behavior?

C=Outcome/Consequence. Find out what happens after the behavior occurs? **WHY?**

\[\text{A} \rightarrow \text{B} \rightarrow \text{C}\]
Flip the model

with permission from Karen Riley, PhD
Determining the Antecedents

- It is sometimes difficult to define the antecedents.
- It is difficult to know where to start.
- We often create additional issues through our attempts to make things better.
- When we make changes in a systematic way we can decrease the need for subsequent interventions.
Goals of Functional Analysis

- Operational Definition of the Behavior
- Prediction of times and situations when the behavior will and will not occur
- Definition of the function (the maintaining reinforcers) that the undesirable behavior produces for the person
Creating a Sound Behavior Plan

• Identify one specific behavior to target.
• Determine when, where and how often the behavior occurs.
• Examine and address any physiologic causes.
• Examine and modify the structure of the situation, when possible to decrease the opportunity for the behavior to occur.
Creating a Sound Behavior Plan

- Provide the child with additional resources for dealing with the stressors associated with the behavior.
- Outline natural consequences for the behavior.
- Be consistent.
- Provide a substitute behavior.
- Reinforce positive behavior.
- Start in a controlled setting.
- Provide opportunities for both success and for failure
Why do most behavior plans fail?

- They assume behavior occurs in isolation.
- They are not holistic in their approach.
- They are too complicated.
- Do not address the foundations of the behavior.
We would never make a sticker chart to help people with visual impairments see better.
We Would Use Visuals to Sequence a Behavioral Intervention

**Hair Pull Protocol**

- Move students
- Don't talk to us
- ONLY come if asked
Use Visuals to Teach Regulation

Don't hurt people

When I'm upset I may hurt people.
Pulling hair hurts.
Scratching hurts.
Kicking hurts.

I will try to be in control of my body.

Ask for a break.
Squeeze hands.
Head on desk.
Go to sensory room.
Take deep breath.
Make Regulation Tools Accessible
Using Visuals to Teach Advocacy
A Behavior Plan Must Consider the Cause or Antecedent of the Behavioral Cycle (previous slide)

- Creating a behavior chart to keep hands “quiet” will fail
- Providing a DRO- differential reinforcement of other (behavior) may work if it targets an effective replacement behavior
- Offering a more adaptive remedy to provide deep pressure WILL work!
• Why are transitions so difficult? The obvious reason relates to the anxiety provoked by the unknown - what is the expectation when I enter the new environment? What is the environment like? Will it be noisy, and chaotic? Who will I see there?
Modify the schedule to assist in transitioning

- It is much easier to modify the schedule then to modify the individual to fit the schedule.
- What are the challenging times? And what are challenging activities? Don’t mix the two.
- What is the purpose of the current schedule and what can be shifted?
- Use physical props to help with the schedule/transitions.
Birth to Preschool

- There is a good deal of evidence about effective interventions for this age group. Early intervention is critical to overall prognosis.
Birth to Preschool
Most Common Issues

- Self-regulation skills begin to rapidly during the preschool age period
- Self-regulation involves the ability to control impulses and expressions of emotion
- Children with difficulties in self-regulation might show a range of problems including higher rates of tantrums, irritable mood, compositionality and disturbances in sleep, eating, activity and attention. (Patterson, 1982, Shaw & Bell, 1993)
Birth to Preschool
Most Common Issues

- Eating (sensory dysfunction) causes the baby to have difficulty tolerating texture and taste which continues throughout the life span.
- This presents behavioral issues as the parent tries to satisfy and nourish the baby.
- Sleep is often disturbed due to hyper arousal and acute auditory sensations.
- Independent toileting is delayed due to neurobiological functions such as textural sensitivity and slow development of sphincter muscle.
Birth to Preschool
Most Common Issues

- Separation Anxiety - difficult to be away from mom and dad
- Language delays both receptive and expressive contribute to the lack of understanding of directions and social language
- Cognitive delays contribute to learning difficulties in preschool
Birth to Preschool
Most Common Behavioral Issues

Fear creates anxiety

Anxiety prompts behavior

Parent has trouble managing behavior

Parent avoids activity

New social activity
Birth to Preschool
Most Common Behavioral Issues

• Lack of communication causes the child to communicate through his behavior
• Sensory dysfunction causes the child to get sensory needs met through maladaptive measures (chewing his clothing, biting his hand, etc.)
Speech/language delays affect behavior

- Difficulty communicating needs which likely results in showing an unmet need through a behavioral sequence such as this:

  The child says I want a drink
  Mom misunderstands and gives him a biscuit
  He gets angry and hits his mom
Visual Supports for Communication Delays

I want

- big wheel
- golf
- trampoline
- bubble blower
- sand and water table
- sidewalk chalk
Disruptive Behavior

• Most frequent type of problem behavior in early childhood
• Disruptive problems often become more prominent in 2 and 3 year olds as rules for compliance and parental expectations increase.
• Tremblay (1998) reported that by age 17 months, 70% of children take toys away from others, 46% push others to obtain what they want and 21-27% engage in one or more of the following; biting, kicking, fighting or physically attacking another child.
• About 50% of oppositional 3 year olds continue to have problems into school age (Campbell, Shaw, & Gilliom, 2000)
Behavioral Problems

When is it significant?

- Rate of behavioral episodes
- Intensity of episodes
- Time to return to baseline
- Specific environmental factors that disproportionately affect the behavior:
  - Transitions
  - Noises
  - Textures
  - Scheduling changes
  - Interruptions in routine
  - Group size
  - Different instructors, teachers, or caregivers
School Aged

Most Common Issues

• Riding to and from school on bus
• Transitioning from home-school-home
• Waiting
• Seeing own image in mirror or window glass
• Hearing or having “Happy Birthday” sung
• Seeing significantly handicapped people
• Hearing screams, conflictual disagreements
• Continuation of behavioral issues diagnosed at preschool age
Social Expectations

• If you are anxious in social settings, you avoid them. When you avoid you become less social.
• It is easier to be with adults than age similar peers because adults repair social interaction and help support successful social collaboration.
• If you have social deficits you dislike talking on the phone, attending social events, being with people you don’t know, and taking social risks.
Social Expectations

Who is your friend?
Sensory Integration Dysfunction

• As the child progresses through school, the environment grows and the child is required to function within a variety of environments
• As the environment grows so do the expectations and norms
• There are a number of ways that sensory dysfunction can contribute to behavioral issues
• Sensory diets, accommodations in the classroom and regulation protocols should be considered when addressing behavioral issues
Our 7 Senses
Sensory Processing and Learning

Figure 5. Pyramid of Learning. (Williams & Shellenberger, 1-4)
So, why is it important?

RESPONSE
A response is generated

PROCESSING
Sensory information is organised and interpreted, stored and related to previous experiences.

SENSORY INPUT
Sensory receptors are stimulated.
Remedies for Sensory Deficits

- Visual symbols for sensory diets
- Provide visual supports for breaks, alternative activities and self regulation
Cool things can be used therapeutically

- Tight fitting clothing can provide deep pressure
- iPhones or pods can provide a distraction or visual schedules
Anxiety

- Sit on chair
- Feet on floor
- Fold hands
- Take 3 deep breaths
- Count to 10
- Good work
Adults

• If you have social deficits you dislike talking on the phone, attending social events, being with people you don’t know, and taking social risks.
• Cognitive deficits limit work choices and opportunities
• Finding the right placement will enhance success
Aging in FX Males

- Often as the adults get older, they are included in fewer activities and can become agoraphobic.
- There is a subset of this population that becomes more agitated, less tolerant and aggressive as they age.
- Psychiatric disorders are common and often result in significant anxiety and phobias.
Characteristics that Impact Work

- Executive function deficit
  - Planning: Coordinating and retrieving information, motor planning (trouble getting started)
  - Inhibition: Modulating/regulating incoming sensory stimuli, concentration/focus (absorbed in own thoughts)
  - Mental Flexibility: Bogged down with details, rigid cognitive set, perfectionism, perseveration (getting stuck)
- Perception (auditory discrimination)
- Auditory processing < Visual processing
- Sequential Processing < Simultaneous processing
- Expressive language/pragmatics
Characteristics that Impact Work

Planning and Execution

1. Difficulty starting or sequencing a task
2. Modulating sensory input - things in the environment are distracting and the reaction is often maladaptive
3. Rigid thinking; inability to remain flexible and problem solve
The Process

This curriculum blends the clients’ interests and skill set with specific job skills. This match enables the client to work productively and independently. The leading cause for failure among DD clients in the work force is boredom and loss of work momentum. Allowing for a better match enables employers to utilize their staff to train and supervise less. If the interest is inherent in the job tasks it requires less coaching and coercion to get the job done. The client is internally motivated to complete the tasks. If there is a discrepancy between the tasks the client is capable of performing and his interests, the use of task analysis to diagnose work strengths enables a better fit to determine what resources are necessary to build employee viability.
Job Coaching

Level I

The client is observed by the job coach in a work rotation so that the coach can analyze how the client performs a variety of work experiences. In addition, the coach assesses the environment to identify what distractors, noises proximity and other environmental factors that may interfere with overall performance and job success. Further, the job coach assesses what aspects of the job may promote work motivation (social contacts, opportunities to move, high interest work content, natural supports in place).
Job Coaching
Level II

The coach’s assessment is used to construct the job elements that best fit the needs of the client. The client is placed in the work environment. The job is then broken into steps with a baseline taken for individual performance on each step. The coach also analyzes the amount of support necessary for each step in order to perform the overall job.
Job Coaching
Level III

After the client is placed in a job and making progress to independence, the level of support for each job is calculated. As the client gains independence needing less and less support from the coach, the natural supports - supervisors and coworkers begin to assume the responsibility of coaching the client to succeed in the work environment and the job coach is faded. If the job needs significant support at this level, decisions about the job efficacy should be made.

The client is assessed using a step by step analysis. Those steps form the basis for data collection using a support matrix to determine how much support is necessary from the trained job coach and when or if the transition to a natural support (job supervisor employed by the site) is possible. Examples of the steps to successful employment follow.
Visual Schedule
Unloading/Loading Produce

- Molly
- Linda
- Marcia
**Outcome:**

**LYFE kitchen**

<table>
<thead>
<tr>
<th>Cue</th>
<th>Response</th>
</tr>
</thead>
<tbody>
<tr>
<td>X sees produce visual on schedule.</td>
<td>X takes the black marker from the wall and moves a black cart next to the boxes of produce.</td>
</tr>
<tr>
<td>Boxes of produce are stacked up.</td>
<td>X uses the marker to write the date on each produce box.</td>
</tr>
<tr>
<td>The date has been written on each box.</td>
<td>X loads boxes onto the cart.</td>
</tr>
<tr>
<td>The cart is full.</td>
<td>X pushes the cart to the cooler.</td>
</tr>
<tr>
<td>The cart is full and in the cooler.</td>
<td>X determines what is in each box and locates where it goes on shelves.</td>
</tr>
<tr>
<td>Location is determined and old produce is removed and put aside.</td>
<td>New produce is placed in back and the old produce is rotated to the front of the shelf with date facing out. X flattens any empty boxes and puts on bottom of cart.</td>
</tr>
<tr>
<td>The black cart is filled with empty boxes.</td>
<td>X wheels out the cart and puts empty boxes by the door. again dates the boxes, fills cart with any remaining boxes.</td>
</tr>
<tr>
<td>The cart is empty.</td>
<td>X locates remaining produce and dates boxes, loads cart, wheels the cart to the cooler and puts away rest of the boxes. X flattens boxes.</td>
</tr>
<tr>
<td>The cart is filled with empty boxes.</td>
<td>X wheels the cart out of the cooler, puts boxes by door, and does not find any more produce to put away.</td>
</tr>
<tr>
<td>There are no more boxes of produce to put away.</td>
<td>X returns the cart to where he found it.</td>
</tr>
<tr>
<td>The cart has been returned.</td>
<td>X returns the marker to the wall. X checks his visual schedule.</td>
</tr>
</tbody>
</table>

**Data Codes:**

NA = Not Applicable
0 = No complaints
1 = Physical prompts for part of task
2 = Gestural prompts for part
3 = Verbal
4 = Visual prompts
5 = Independent no prompts
<table>
<thead>
<tr>
<th>Job: Tortilla Chips</th>
<th>Date of Probe</th>
<th>Data code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cue</strong></td>
<td><strong>Response</strong></td>
<td></td>
</tr>
<tr>
<td>Food prep is on the visual schedule</td>
<td>D. checks in w/ the kitchen staff</td>
<td></td>
</tr>
<tr>
<td>Tray needs parchment paper</td>
<td>D. places parchment paper on tray</td>
<td></td>
</tr>
<tr>
<td>Paper needs to be sprayed w/ no stick oil</td>
<td>D. Sprays the tray w/ oil</td>
<td></td>
</tr>
<tr>
<td>Spray is evenly spread on sheet</td>
<td>D. holds spray bottle above the paper</td>
<td></td>
</tr>
<tr>
<td>Tray is prepped and tortillas cut</td>
<td>D. places cut tortillas onto the tray</td>
<td></td>
</tr>
<tr>
<td>Tortillas are on tray</td>
<td>D. holds spray bottle above the tortillas and sprays</td>
<td></td>
</tr>
<tr>
<td>Shaker w/ seasonings is pre cut</td>
<td>D. picks up shaker and shakes evenly onto tortillas</td>
<td></td>
</tr>
<tr>
<td>Tray is filled w/ tortillas</td>
<td>D. takes tray and places on rack</td>
<td></td>
</tr>
<tr>
<td></td>
<td>D. checks his schedule</td>
<td></td>
</tr>
</tbody>
</table>

*Data Codes:*
- NA = Not Applicable
- 0 = No response
- 1 = Physical prompts for all or most
- 2 = Physical prompts for part
- 3 = Verbal or gestural prompts
- 4 = No added prompts
Food Prep

![Chart showing food prep tasks for different days and staff members between 7/23/2014 and 8/27/2014. The chart includes tasks labeled 'Molly', 'Linda', and 'Marcia'.]
The Not So Scientific Study

Taken from National FX Foundation’s Adolescent and Adult Project

Several parents and professionals came together to interview and survey parents with children with Fragile X Syndrome over a year’s time

They provided the information to the foundation who then produced a book with DVD to transmit the information

Marcia L. Braden, PhD
Predictors of Independence

- Encourage a variety of experiences within the home community
- Develop interests and “common denominators” that make them more viable social partners
- Encourage social engagement in small groups such as church activities, scouting or clubs
- Encourage age appropriate dress, interests and activities
- Encourage grooming and personal hygiene

Marcia L. Braden, PhD
Behavioral Issues In Adolescents and Adults

- Mood Disorders
  - Unable to sit for long
  - Paces aimlessly
  - Sleeplessness
  - Irritability

- Depression
  - Loss of interest in favorite activities
  - Lack of appetite
  - Increased rigidity in routines
  - Nervousness

- Panic Episode
  - Become flushed
  - Look frightened
  - Try to flee
  - Breathe rapidly
  - Sweat heavily
  - Rapid pulse rate
  - React with self-injury

Taken from National FX Foundation’s Adolescent and Adult Project
Socialization

Social venues that have brought positive experiences to those with FXS

- Church
- Sports/Special Olympics
- Theatre/Movies
- Karate
- Choir
- Boy/Girl Scouts

Taken from National FX Foundation’s Adolescent and Adult Project
Teach Prosocial Behavior

- Discriminate between appropriate and inappropriate behavior

- unzipping pants in public
- exposing private parts in public
- using sexually explicit language in public

Marcia L. Braden, PhD
Teach Prosocial Behavior

- Teach basic concepts of Private and Public Behavior, Places and People
- begin early
- model appropriate sexual behaviors
- use repetition to habituate appropriate behaviors
- teach advocacy and self defense
- participation in social skills groups

Marcia L. Braden, PhD
Teach Prosocial Behavior

Private

Private

Public

Public

Public

Public
Social Compass Program

Marcia L. Braden, PhD
The Social Compass Program

• Four Components
  ■ Charting Social Targets: Meeting, Greeting and Keeping
  ■ Mapping Out Manners
  ■ Navigating Sexuality
  ■ The Compass Club

Marcia L. Braden, PhD
The Social Compass Program

• *Charting Social Targets; Meeting, Greeting and Keeping*
  ● This component teaches a sequence of social skills to be taught in a developmental spectrum
  ● Teaches specific skills related to social interaction so that friendships can be developed

Marcia L. Braden, PhD
The Social Compass Program

• *Mapping Out Manners*

  This component includes the teaching of good manners and respect

  Behavioral targets are grouped into domains and divided into two levels that are age-linked

  This component includes a DVD of video vignettes to demonstrate the contrast between good and poor manners

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The Social Compass Program

• *Navigating Sexuality*
  Includes a developmental model of sexuality to include age appropriate sexual behavior

This component includes a DVD of video vignettes with instructional objectives to prompt frank discussion about the DVD

Marcia L. Braden, PhD
The Social Compass Program

- *The Compass Club*

This component includes a collection of games and activities that can be used to reinforce the concepts taught through the other components.

These activities can be used in groups or on an individual basis.
Social Compass Program
Level 1 spinner and cards
Level 2 spinner and cards
Social Compass Teaching Boards
RESOURCES

- Fitting In and Having Fun, Vol. 2: Moving on to Middle School – www.difflearn.com
- The National Fragile X Foundation’s Adolescent and Adult Project: Smoothing the Transition to Successful Adulthood
- Community Success CD
- Conversation Skills On the Job and in the Community
- Social Skills at Work CD
- Social Skills on the Job CD set
  www.AttainmentCompany.com
Case Study GV

• Public school placement was not meaningful and much of his time was spent engaged in non functional tasks

• His behavior was controlled using contingencies and response cost techniques…
  “if you do X, you will lose your video”

• As expectations increased he was less willing to comply and protested through his behavioral outbursts

Marcia L. Braden, Ph.D.
Case Study GV

• In HS his behavioral episodes became so violent he could not ride a school bus, attend full day or attend community outings
• The school district could no longer educate him in a public school setting and developed a school program to be implemented in his home
• A team of behaviorists worked in the home to teach compliance behavior
The team increased their intervention time and expectations.
High interest materials were used to hold his attention and increase motivation.
Visual systems, choice boards and frequent reinforcement increased participation and decreased avoidance behavior.
Removal of negative attention helped decrease physical aggression.
Case Study GV
Home Routine

• Picture schedule of morning routine with photos of GV showering, dressing, eating breakfast, brushing teeth, packing up backpack and getting ready to get on the school bus.

• This routine was planned carefully to avoid a long wait for the bus which became a trigger for aggression
Transition Data
Case Study GV

- Teachers from The Joshua School, a private school for individuals with autism cross trained with the home team.
- The home team taught him a morning routine; shower, dressing, eating breakfast and getting ready to transition from home to bus.
- Home behaviorists co taught with teachers from the private school.
- He attended school on a reduced schedule.
Behavioral Supports

- Use of visual support systems across environments and activities
- Providing forced choices in daily schedule
- Support person who has strong bond is consistent and can read his moods.
- Good communication between providers and parents
- Variety of materials and academic methodologies
- Provide sensory input/opportunities/options

Marcia L. Braden, Ph.D.
Case Study GV

• Gradually time was added to his school day and he was able to attend full day after one year
• He is now attending full day, attends all school functions and community based instruction
• His aggressive behavior has been reduced significantly and is no longer “at risk”

Marcia L. Braden, Ph.D.
Afternoon Session
Start with the Environment

• The environment is the easiest thing to change.
• The environment can have a huge impact on behavior as the neurobiology collides with the environment.
• Changing the environment can shed light on the antecedent of behaviors.
• The environment is anything physical.
It is difficult to

Overcome this

With this
Or this..with this
Creating a Good Place to Learn

- Consider lighting, visual clutter and extraneous noises
- Utilize visual supports whenever possible
- Reduce verbiage
- Organize spaces with desk placement and work table charts
Creating a Good Place to Learn

• Visual timetables
• Consistent routines that build predictability
• Boundaries
• Movement breaks (www.yourtherapysource.com)
• Heavy work activities
• Fidgets
• Seating options
• Fast paced presentation
• High interest materials to enhance engagement
• Opportunities to interact socially with adults and peers
Use physical props to help control the aspects of the environment that you can’t control.
A Good Place to Learn

• Environment
• Instructional Quality
• Structure of the Classroom
• Visual Supports and Structure
• Sensory Integration and Regulation Protocols
• Opportunities for Inclusion with Typical Peers
• Opportunities to cooperate and help in the classroom
A Good Place to Learn

• Incorporate high interest material into instruction
• High interest links familiarity with novelty
• Novel skills are not taught in isolation, but embedded into the high interest materials
• Interests may neutralize the full impact of learning something new and unfamiliar, hopefully reducing anxiety
• Utilize peers as teaching models. This facilitates indirect learning and reduces the intensity of the learning experience.
• Use side dialoging to introduce a schedule change, subtle shift in activity or other communication need.
Cognitive Phenotype and Educational Strategies
Cognitive Profile

• Strengths
  Vocabulary in context
  Long term memory for high interest, meaningful information
  Face emotion

• Weaknesses
  Attentional control and dysregulation
  Linguistic processing
  Visual motor impairment /motor planning
  Lower IQ scores
  Executive functioning
Summary of Cognitive Research

- Strengths in vocabulary (Dykens et al., 1989), long term memory for meaningful and learned information (Freund and Reiss, 1991) and face emotion perception (Turk and Cornish, 1998).
- Weaknesses in attentional control (Munir et al., 2000), linguistic processing (Belser and Sudhalter, 2001), and visual spatial cognition (Cornish et al., 1998, 1999).
• Visual-motor impairments for tasks that require drawing skills (Crowe and Hay, 1990; Freund and Reiss 1991) Impairment in tasks requiring psychomotor coordination such as pegboard (Cornish et al., 1999)
• Both biological (amount of protein produced) and environmental factors predict outcomes of children with full mutation (Dyer-Friedman et al., 2002)
SUMMARY OF COGNITIVE RESEARCH

• FMRP depletion was strongly correlated with lower IQ scores in males with FXS (Loesch, et. Al., 2003)
• FMRP depletion was related to slow processing speed, poor short-term memory and attentional dysregulation (Loesch, et. Al., 2004)
Measuring IQ Over Time

• Early studies reported a decline in IQ over time. However, a number of methodological problems were noted in the studies. For example, insufficient sampling and behavioral deficits were not taken into account (Hay 1994)

• A retrospective study done by Wright-Talamante in 1996, indicated that the decline was more likely due to a lack of cognitive development rather than any type of degeneration of the CNS
Cognitive Phenotype
Studies of IQ in Males with FXS

Kemper et al. 1988
K-ABC  N = 20  ages 4-12

1. FRA-X IQ < than DD controls
2. FRA-X Achievement score > DD Controls
3. FRA-X has more variation across subtests than DD controls
4. FRA-X Simultaneous (mean 71) > Sequential (mean 62)
Cognitive Phenotype

• Sequential vs. Simultaneous
  • Sequential A-B-C-D – the parts
  • Simultaneous ABCD – the whole
Simultaneous Processing

- Gestalt-like configuration
- Global conception
- Intuitive method of organization
- Multiple stimuli
- Visual memory for the “whole” is better than the parts
# Build On Cognitive Strengths To Address Weaknesses

<table>
<thead>
<tr>
<th><strong>Strengths</strong></th>
<th><strong>Weaknesses</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>• Long-term Memory</td>
<td>• Need for Closure</td>
</tr>
<tr>
<td>• Associative Learning Preferences</td>
<td>• Focus and Concentration Deficits</td>
</tr>
<tr>
<td>• Indirect Instruction</td>
<td>• Processing Deficits include auditory processing which may have been caused by recurrent ear infections</td>
</tr>
<tr>
<td>• Responds to visual cues</td>
<td>• Executive Functioning Deficits</td>
</tr>
<tr>
<td></td>
<td>• Inconsistent Reaction to Stimuli – Sensory Integration Dysfunction</td>
</tr>
<tr>
<td></td>
<td>• Difficulty Generalizing – Learning life skills are best taught within a functional context.</td>
</tr>
</tbody>
</table>
Executive Function

What are Executive Functions? (Papolos & Papolos, 2002)

• The most advanced and complex functions of the brain
• Linked to intentionality, purposefulness and complex decision making
Executive Function

- Executive Function is required whenever going “on automatic” would not be sufficient and especially when it would lead one astray (Adele Diamond, PhD.)
- The ability to direct and switch attention, inhibit repetitious behavior, and inhibit appropriate responses (Wilding, Cornish, and Munir 2002)
Ways to remediate EFD

- Rhythm
- Music
- Do the first step for them
- Fill-ins
- Backward chaining
- Change intonation patterns
- Use an accent
Fragile X Syndrome & Sensory Processing

Individual’s with fragile x syndrome are frequently over sensitive to many types of sensory input. This over sensitivity makes being in groups and being in the community difficult. It may also contribute behavioral outbursts.
Visual supports to aid in learning

Crossing off tasks is reinforcing

Sensory choices assist in establishing self regulation
Visual Supports Build Social Language and Spontaneous Communication
Strategies to Build social skills

Conversation Ladders
A Good Place to Learn

- Structured Teaching uses gestalt staging. This is consistent with seeing the whole and not the parts. Students with cognitive delays often require a concrete presentation.
- Non sequential learners can benefit from a global visual picture which includes the “whole” of what is expected.
- Provide the entire sequence of a task so that the student can see the end to better understand the completion and steps to get there.
- Encourage flexibility in responding- help the student adapt to new situations.
A Good Place to Learn

Customized token boards provide clear expectations, summons attention and provides motivation.
A Good Place to Learn

• Structured teaching provides appropriate and meaningful environments that reduce stress and anxiety.
• Visual backups provide support when the student is unable to receive meaningful benefit from traditional instruction.
• Visual support is always there to refer to whenever frustration mounts.
• Rules and consequences are clearly defined reducing the need to manipulate or “deal”.
• Expectations are clearly defined making it easier to understand and ultimately comply.
A Good Place to Learn

Provide visual reminders for behavioral programming
A Good Place to Learn

First

Then

First → Then → Next

Image: "First" and "Then" sections with images of '2+4=6' and 'ball bounce'. Another image showing a visual aid for "First Goes To Playground" with images of a boy, girl, car, and a playground.
A Good Place to Learn

- Structured teaching provides concrete parameters and physical organization of the learning environment
- Provides specific guidelines and expectations to promote the development of independence and task completion
- Provides limited input, applying reduced verbal input with visual supports. Helps the student focus on the concept and not the details.
- Consistency of expectations provides predictability and takes out the guess work
A Good Place to Learn

- Providing choices between activities gives students a sense of control
A Good Place to Learn

- Structured teaching provides predictability and sameness which is calming and stabilizing to students.
- Anxious students may engage in flight or fight behaviors, preoccupations, perseveration, obsessions (the thought) and compulsions (the behavior) which can be reduced with predictability and clear expectations.
- Clearly defined expectations, assignments and time lines are comforting and allow the student who is anxious to access their full potential.
A Good Place to Learn

- Students with FXS often have difficulty interpreting sensory input and need an area in which to learn how to self-regulate.
A Good Place to Learn

Indicate changes by placing a symbol/word by the activity
Visual supports to aid in learning

- Object schedule
- Dry erase board
- 3-ring binder
- Clip board
- Manila folder
- Written list schedule
- Sentence schedule
- Photograph schedule
- Picture/Symbol schedule
- Picture/Symbol with written schedule

With permission from Julie Race and Jody Minarick
Symons, Clark & Roberts 2001

- Classroom engagement of elementary school children with FXS is strongly related to the environmental and instructional quality of the teachers and classroom.
- The ways the teachers structured and arranged the classroom environment was much more important to student engagement than specific aspects of the child’s FX status, medication use or dual diagnosis.
Symons, Clark & Roberts (2001)
What does it mean to you?

• Classroom structure- be sure the student has access to the focus of instruction

• Remember learning strengths and weaknesses:
  IF YOU TEACH TO THE WEAKNESSES YOU WILL GET LOW FUNCTIONING LEARNERS AS WELL AS MORE BEHAVIORAL INTERFERENCE

• Include more visual supports and less verbiage
Symons, Clark & Roberts (2001)  
What does it mean to you?

- Avoid sequential instruction; if the task is broken down, allow the student to execute the task using his strengths (simultaneous)
Encouragement

- Children are sensitive
- Look for small improvements
- Give a great deal of praise
- Increase your level of enthusiasm and excitement
- As time goes on require larger gains for encouragement
- Pay attention to signs of frustration and anger
- Back away when your child becomes agitated
- Provide a break and return to the task at hand
- Always end on a positive note
Strategies and Methods
to teach math, reading and writing
THE DIFFICULTY WITH MATH
Math

Why is math so difficult?
• Math is sequential
• Math builds on a sequence and cannot
  be taught through a context like reading
• Many aspects of math require memory
  for unrelated facts
• Teaching 1:1 correspondence is compromised by
  poor impulse control and slowing down in order to
  count objects on at a time
Math

Use patterns to teach math order and the gestalt of a number sequence
Math

• Teach math sequence incidentally
• Use dot math, matching and number lines
• Utilize patterns to teach
Math
Math

Continue to build an understanding of number value by matching an amount to a numeral
Math-Equivalence Board

- Based on the "stimulus equivalence" paradigm
- Equivalence and = is akin to "put with same"
- Can be expanded to teach content information through associations and equivalence such as: digital and analog time, money and digital equivalence, 1:1 correspondence/object to numeral
Math-Equivalence Board
Math-Equivalence Board
Math-Equivalence Board
Math
Math

- Using money to understand counting can be effective especially when there is a purchase used as a reward.
- Token boards can use pennies, nickels, dimes and quarters
- Tokens (coins) can be used to buy things at a school store, treasure chest or vending machine
Math

The use of the penny board teaches the student to earn tokens (money) to buy a reinforcer
Math

Placing coins on a configured pattern teaches the number value
Math
Math
Math

• Use closure strategies
  \[5 \_ 7 \_ \_ 10\] or \[6 + \_ = 9,\] \[3 + \_ = 9,\] \[6 + 3 = \_\]

• Use real life experiences to make math functional
  calculator, computer programs, purchases, microwave
Math

- Pairing a reinforcer with the price of purchase helps teach money identification as well as the value
Math

Understanding math requires repetition.
Vary the materials to teach the same concept
Understanding math requires repetition. Vary the materials to teach the same concept.
Math
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Understanding math requires repetition. Vary the materials to teach the same concept.
Errorless Learning

- Do not allow your child/student to make mistakes as they begin to learn words
  - guide your child’s/student’s hand so they always make a correct match
  - adjust the difficulty level of a task to be sure your child/student is successful
- Ensures initial success which leads to greater feelings of competence and motivation
The Edmark Reading System uses a visual format to teach comprehension.
Reading

The use of a visual-picture system such as rebus or Mayer Johnson provides visual support for language comprehension.
Use objects 
To build an understanding of the words
LOGO READING SYSTEM- Braden
Who Can Benefit?

Non readers from the ages of 3-

Students who have failed to learn to read using other methods

Students who need high interest materials to become engaged in a structured reading system
LOGO READING SYSTEM-Braden

- Incidentally Acquired Skills
- Fade out logo
- Retain text font
- Place words with text font into phrases
- Rhyme words with target words
  hut/shut/cut/nut, etc.
Word Families

- Children with FXS remember information within a whole, not within parts

- Using parts of words or word families enables the student to read more words by adding first letters to word families

- For example the word family *and*
  - B-and = Band   H-and = Hand   S-and = Sand
WORD BUILDER-BRADEN
WORD BUILDER-BRADEN

Pairing a picture with a word uses the visual strengths of those with FXS
WORD BUILDER - BRADEN
Reading

- Use high strength interests to build word recognition
Reading
Reading
Why Make It?

Interests + Learning Style = Success

Traditional teaching methods have proven unsuccessful

Why continue to push a square peg into a round hole?

How many times is the avoidance behavior due to the fact that the materials are boring without an association or relevance?
Reading

• Matching
  • Symbol to symbol (token boards)
  • Picture to picture
  • Word to picture
  • Phrase to picture
  • Sentences to test comprehension
High interest matching
Matching picture to word
Matching picture to category
PHRASE TO PICTURE

- Her friend is a
- She eats a poison
- She lives with dwarfs
- She wears glass slippers
- She loves a beast
- She has a horse named
FILL-IN USING HIGH INTEREST MATERIALS

Turner is a flat-head ____________________________.

   hammer          screwdriver          wrench

Manny is a ____________________________.

   carpenter       girl           dog

Pat is a ____________________________.

   wrench         saw           hammer

The monkey wrench is named ________________.

Rusty   Pat    Manny
SENTENCES TO CHECK FOR COMPREHENSION

I want the letter puzzle

I want to play a game
TASKS TO CHECK FOR COMPREHENSION

- 2 Red Buses
- 1 Green Airplane
- 3 Red Fire Trucks

- 1 Purple Car
- 2 Blue Fire Trucks
- 3 Yellow Boats
TASKS TO CHECK FOR UNDERSTANDING

1 purple bead
3 blue beads
3 orange beads
Steps to Successful Inclusion

- History behind STSI
- If the student is included during the school day what does he/she miss in SPED setting?
- Identify the outcome and reflect it in IEP goals social, behavioral, academic?
- Use probe data to the efficacy
Steps to Successful Inclusion

Started with the STAR curriculum data sheets for functional routines

- Bathroom
- Specials (art, music and PE)
- Small group
- Teacher table
- Lunch
Steps to Successful Inclusion

• The structure is first established in the special education setting
• The instructional supports are provided and taught in a SPED setting
• As the student becomes more independent and needs less instructional support based on the probe data, the structured supports became essential.
• The structure and visual supports are adapted to be used in the general education setting
Steps to Successful Inclusion

- Jobs
  - Sharpen Pencils
  - Calendar
  - Wash Table
  - Wash Chairs
  - Stack Chairs

- Centers
  - Reading Center
  - Math Center
  - Sensory Center
  - Computer
Steps to Successful Inclusion

• As the student becomes integrated into the general education setting, the natural supports provided by the classroom teacher are utilized.

• Those natural supports are surveyed by the SPED teacher to determine if additional supports are necessary.
Support Steps to Successful Inclusion

- Did not argue
- Follow directions
- Listen to teacher
- Answering questions
## Steps to Successful Inclusion

<table>
<thead>
<tr>
<th>Objective</th>
<th>Proficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Attend to teacher at desk or circle/instructional probes</td>
<td>3 5+ 10</td>
</tr>
<tr>
<td>2) Complete task with typical classroom supports</td>
<td>3 5+ 10</td>
</tr>
<tr>
<td>3) Sustain focus on task for time requested/instructional probes</td>
<td>3 5+ 10</td>
</tr>
<tr>
<td>4) Sustain focus on visuals/at desk or circle/instructional probes</td>
<td>3 5+ 10</td>
</tr>
<tr>
<td>Cue</td>
<td>Response</td>
</tr>
<tr>
<td>------------------------------------------</td>
<td>------------------------------------------------------------</td>
</tr>
<tr>
<td>Schedule indicates X</td>
<td>Student goes to designated area in classroom and waits</td>
</tr>
<tr>
<td>Classroom teacher gives instruction</td>
<td>Student attends to classroom teacher</td>
</tr>
<tr>
<td>Academic materials are required</td>
<td>Student gets required materials</td>
</tr>
<tr>
<td>Classroom teacher gives assignment</td>
<td>Student works on assignment appropriately</td>
</tr>
<tr>
<td>Classroom teacher is available</td>
<td>Student asks for help in an appropriate way if needed</td>
</tr>
<tr>
<td>Classroom teacher indicates X is over</td>
<td>Student moves/transitions from the activity</td>
</tr>
<tr>
<td>Data codes</td>
<td>0= No response</td>
</tr>
<tr>
<td></td>
<td>2= Physical prompt for part</td>
</tr>
<tr>
<td></td>
<td>3= Verbal or gestural</td>
</tr>
</tbody>
</table>
QUESTIONS?