

Behavioral Characteristics And Intervention Strategies

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The Neurobiology of Behavior

- Biological breakdown of FX- in 95% of those w/FX there is an expanded mutation meaning that the DNA is stretched out in the area of the promoter region
- This promoter region is an expansion of the nucleotides CGG

The Neurobiology of Behavior

Fragile X Gene FMR 1

Mutable CGG Repeat Region

Normal CGG repeats 50 - 59

Premutation (Carrier) CGG repeats 60-200

Usually unmethylated/clinically unaffected

Full Mutation CGG repeats 200-2000

Methylated/affected 95% males affected

50% females affected

The Neurobiology of Behavior

- The promoter is like the light switch of the gene and when the repeats get too big, the gene is switched off



- So then the gene stops making what it is supposed to make like the RNA (ribonucleic acid). The RNA can't make the protein which is called FMRP

The Neurobiology of Behavior

- FMRP is very important for the development of the brain and necessary for the brain connections. If the connections aren't made, the brain can't work properly
- Glutamate systems are responsible for regulating the FMRP. Glutamate is the most common neuro transmitter

The Neurobiology of Behavior

- The mGluR system (metabotropic glutamate receptor) interacts with the mGluR 5 receptors which activate the brain
- When the receptors get activated, protein synthesis is initiated (this is a complex process)
- FMRP serves as the feedback or inhibition trigger for the brain and the mGluR
- is the accelerator



The Neurobiology of Behavior

- You need a balance between the mGluR and the FMRP because if you don't stop making protein you run the risk of making too much
- In FXS you make too much protein and the brain connections can't form properly
- In some parts of the brain the connections get too strong as in the (amygdala) or the anxiety center

The Neurobiology of Behavior

- And in other areas the connection is too weak like in the Cortex of the brain (hippocampus) which can cause thinking to be impaired and/or affect short-term memory
- Problems in the Frontal Lobe (mGluR) affect hyperactivity

So How Does This Compute to Real Life Behavior Problems?

- Add pictures here

Characteristics that Impact Behavior

- Cognitive deficits
- Hyperactivity, poor impulse control, inattention
- Sensory integration dysfunction
- Speech and language delays
- Gross and fine motor delays
- Physical ailments
- Social anxiety

These characteristics need to be understood, but **CANNOT** be used as an excuse for inappropriate behavior.

- Picture here

Caution!

Some behaviors are -

- Developmentally appropriate
- Part of the phenotype
- An appropriate compromise
- Not worth fighting about



FACTORS THAT AFFECT BEHAVIOR IN THOSE WITH FXS

- **HYPERACTIVITY OFTEN REFERRED TO AS ADHD**
Excessive motion, darting about, rushing, difficulty staying still
- **IMPULSIVITY**
Difficulty waiting until needs can be met; requiring immediate attention, making decisions quickly without forethought.
- **ATTENTION AND CONCENTRATION OFTEN REFERRED TO AS ADD**
Difficulty sustaining attention, easily distracted, unable to focus and attend.

FACTORS THAT AFFECT BEHAVIOR IN THOSE WITH FXS

- **CHANGE IN ROUTINE AND
TRANSITIONS**

Easily upset by changes in schedule, routine and people or perceived expectations.

FACTORS THAT AFFECT BEHAVIOR IN THOSE WITH FXS

- **RITUALISTIC AND REPETITIVE
BEHAVIORS**

Compulsive repetition of hand movements, behavioral rituals such as turning lights off and on, closing doors and drawers and verbal repetitions or perseverations.

FACTORS THAT AFFECT BEHAVIOR IN THOSE WITH FXS

- **SOCIAL ANXIETY/SHYNESS**

Difficulty interacting with others upon request. It appears to be aversive to force eye contact, handshakes or other forms of social interactions.

General Considerations

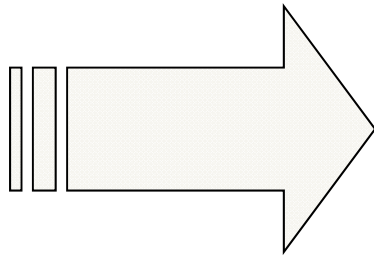
- Holistic Approach
 - Behavior does not occur in isolation
- You may employ several modifications, but only one behavior plan at a time
- Understand why **YOU** are doing what you are doing
- Freedom within structure, CHOICES

Understanding Problem Behavior

- Disruptive to home or school environment
- Interferes with the child's ability to learn
- Interferes with other's ability to learn
- Presents danger to self or to others
- Interferes with social acceptance

Anxiety

Perceived
Danger



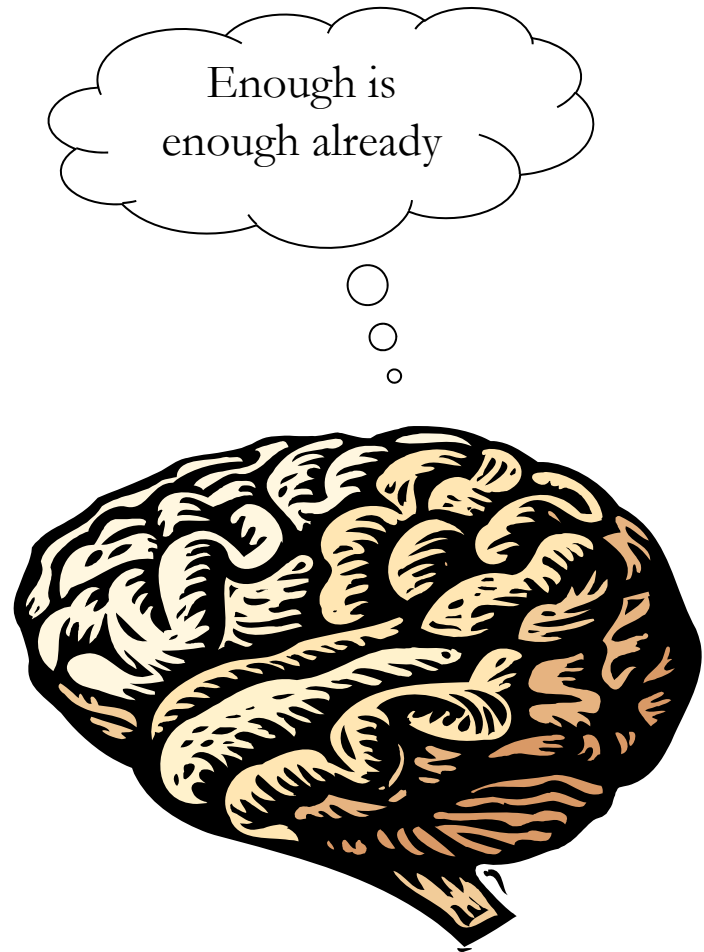
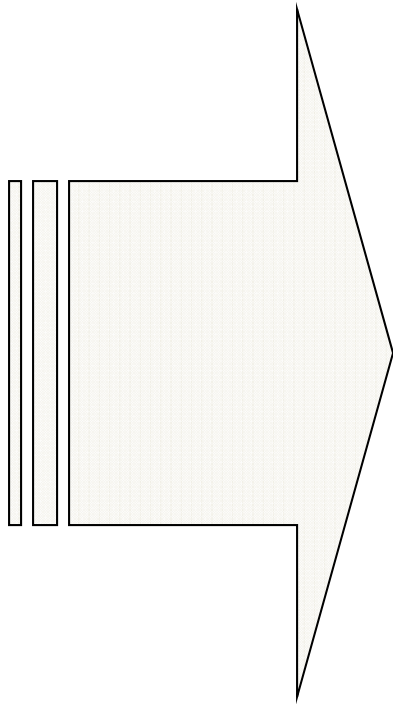
Normal Anxiety-

- A neurobiological response to danger.
- The brain's way of waking up the central nervous system.
- Prepares the body for fighting a threat
 - or flight from a threat.

ANXIETY AND NEGATIVE EMOTIONS

- Anxiety plays an important role in negative emotions such as anger, rage and irritability
- These feelings can result in behavioral episodes such as aggression and explosive outbursts
- Inconsistent regulation of the arousal functions (attention, motor control, and impulses) contribute to a fear of being out of control

Anxiety



Anxiety Perceived Danger

(The Feeling)

- Tension
- Hypervigilance
- Worry
- Fear
- Stomach Ache
- Head Ache
- Racing Heart
- Chest Pain
- Digestive Discomfort

(The Behavior)

- Agitation
- Eye aversion
- Jaw/fist clenching
- Multi-focuses
- Over-reactivity to stimuli
- Sweating
- Muscle Tension
- Nervous picking

Symptoms Associated with Anxiety

- Preoccupations
- Perseveration
- Obsessions (the thought)
- Compulsions (the behavior)

Perceived Threats

- Social
- Verbal
- Sensory
- Motor
- Transitions
- Failures
- Frustration
- Physical

Fear Can Take On A Fight or Flight Reaction

Fight often manifests in the following behaviors:

Argumentative

Oppositional

Relentless

Aggressing

Violent

Rueful

Fear Can Take On A Fight or Flight Reaction

Flight often manifests in the following behaviors

Hyperarousal

Obsessive/Compulsive

Avoidance

Covers eyes, face, plugs ears

Panic

Phobic

SOCIAL ANXIETY - SOCIAL WITHDRAWAL

- Difficulty interacting with others upon request. It appears to be aversive to force eye contact, handshakes, interaction with strangers and exposure to stressful novel events.
- There appears to be a subset of people affected by FXS that may show more persistent avoidant behaviors which may be predictive of high risk for ASD (Hessl et al., 2002, Kaufmann et. al., 2003)

Creating a Sound Behavior Plan

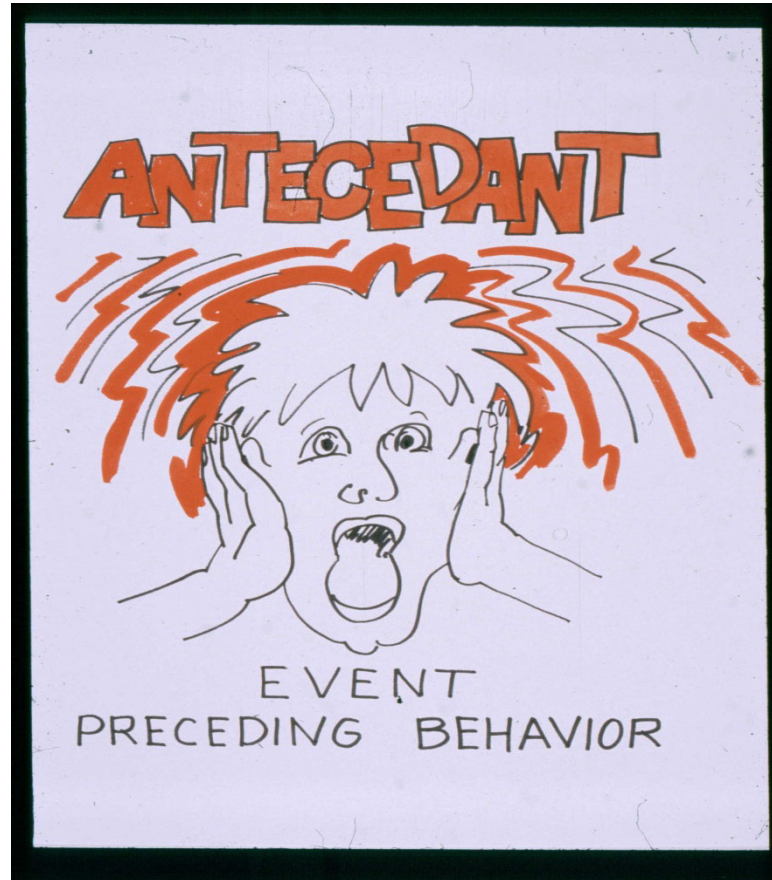
THE ABC'S OF BEHAVIORAL PROGRAMMING

A – Antecedent: What happens before the behavior

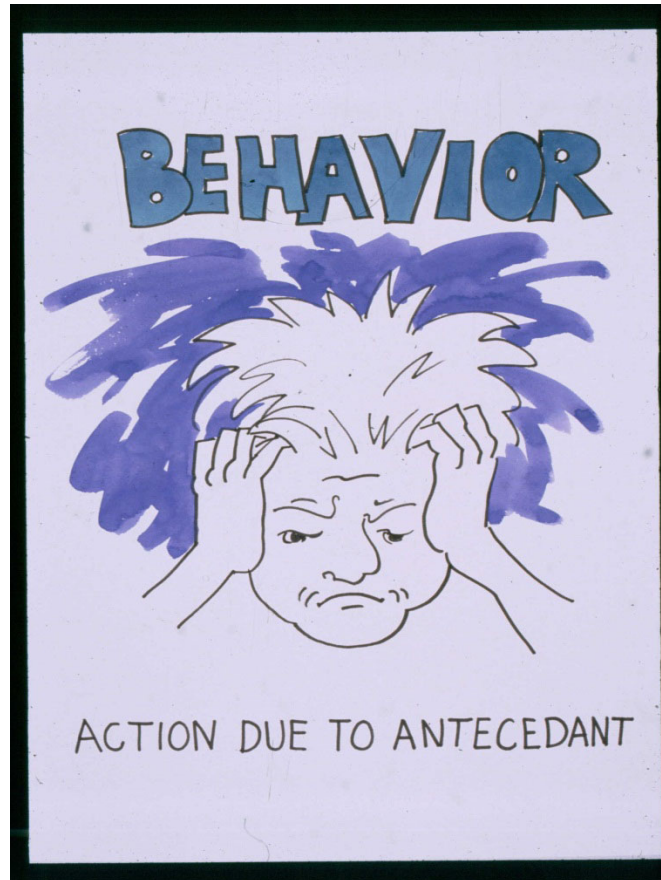
B – Behavior: The overt or covert act

C – Consequence: The outcome

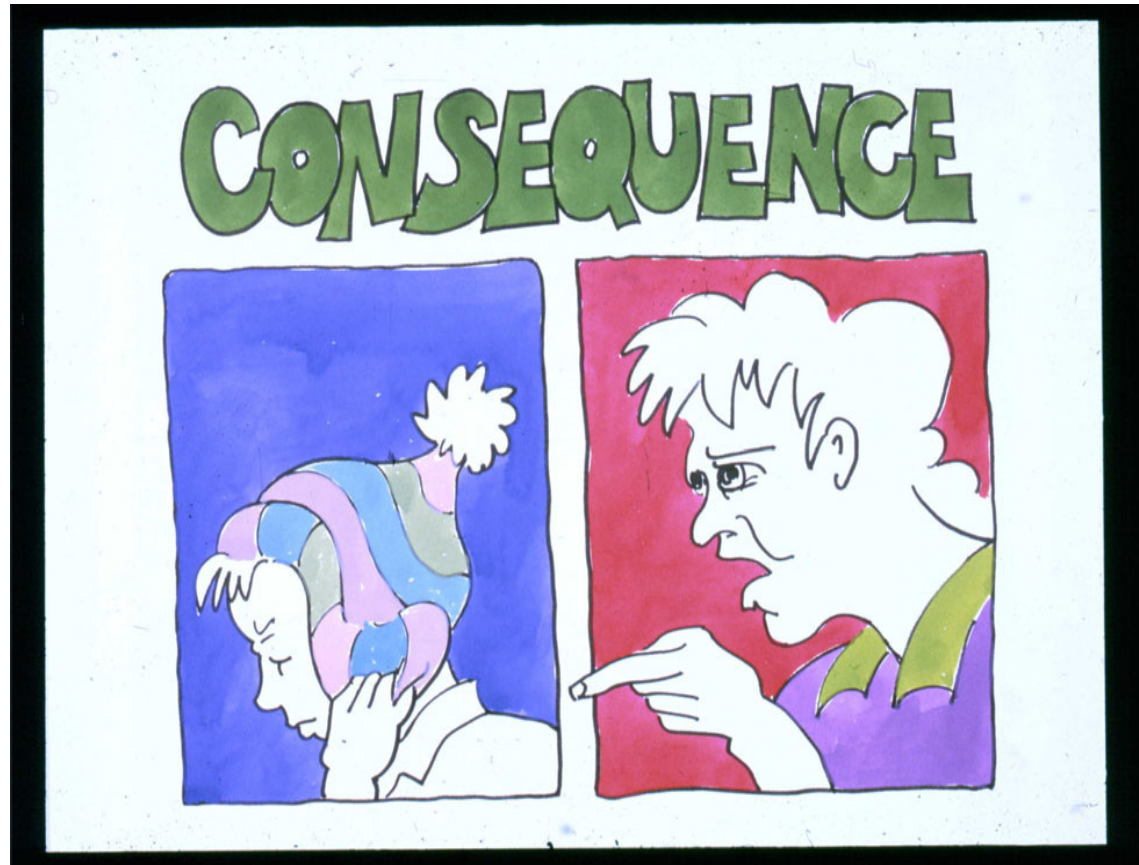
Behavioral Factors



Behavioral Factors



Behavioral Factors



Antecedents

- SOCIAL ANTECEDENTS
- ACADEMIC ANTECEDENTS
- ENVIRONMENTAL ANTECEDENTS
- PHYSICAL ANTECEDENTS

SOCIAL ANTECEDENTS

- Introductions, eye contact, hand shaking
- Perceived or real conflict/confrontation
- Embarrassment, compliments, criticism, attention
- Verbal interaction, phone calls, conversational exchange, direct questioning

Social Antecedents

- Typical defenses to being overstimulated include: dropping to the floor, hiding behind a parent, chewing and mouthing objects and hiding behind hands

Social Antecedents

ENVIRONMENTAL ANTECEDENTS

- Loud, unfamiliar or unpredictable noises
- Transitions, changes in routine
- Artificial, strobe or flickering lights
- Crowded conditions

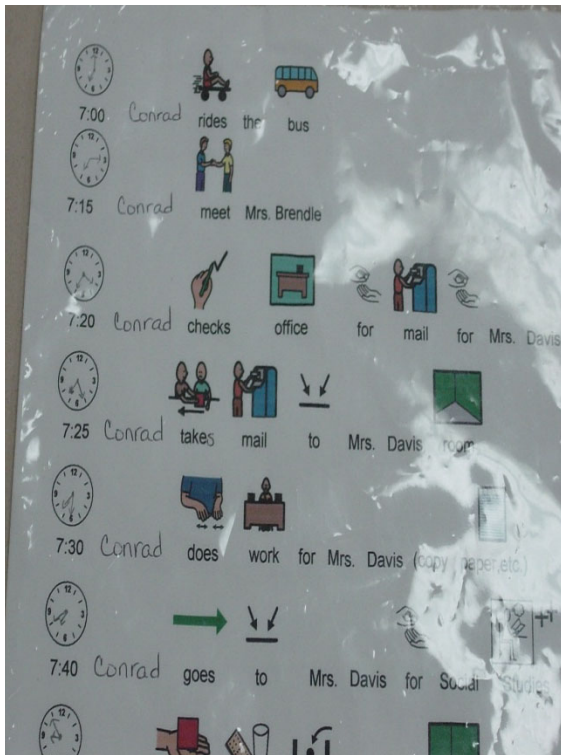
Crowded Conditions

PHYSICAL ANTECEDENTS

- Red ears, face or neck area, quickened heartbeat, trouble breathing, increase in perspiration, tight muscles
- Speech gets faster, louder and more cluttered
- Closing eyes, covering face, turning face, averting gaze
- Hand flapping and stereotypies
- Hyperactivity, increase motor activity, irritability
- Need to escape or avoid

Etiological Components Of A Positive Behavior Support Plan

- Provide predictability of schedule as much as possible



JOBS	NOT DONE
Collect newspapers	None
Clean checklists	Jason
Wipe scuffs off	Tyler
Wash chairs	None
Wash counters	Tyler
Wash tables	None
VACUUM	Jason



Etiological Components Of A Positive Behavior Support Plan



Use of a Visual Schedule

Etiological Components Of A Positive Behavior Support Plan

- Positive reinforcement
 - Tokens used to signal “on task” behavior and compliance to redirection
 - Points or tokens for delayed reinforcement (end of day)
 - Verbal or gestural reinforcement as needed to encourage success

Example Of A Positive Behavior Support Plan

- Curricular adaptations will include high-strength academic materials. Staff will limit verbal input while pairing with visual support.
- Antecedents will be considered before intervention or consequences are applied
- Picture schedule with frequent breaks will be provided on a consistent basis.
- Use social stories to teach more adaptive behavior

Use social stories as a behavioral remedy

Seth loves to go out to eat especially when he can eat his food outside. He likes it outside because there is more room and it is less crowded and noisy. *Insert a photo of Seth eating outside*

Sometimes Seth eats at a restaurant with his mom and dad. This is hard because he can tell when it will be noisy and crowded. When he sees that the parking lot is full, he worries about how crowded the restaurant will be. When he gets inside the restaurant and it is noisy, he says, "I'm scared." This is good because then his parents will know to order food and take it home or eat it outside. *Insert a photo of a restaurant you frequent with Seth where it is usually crowded*

Sometimes Seth gets so scared he forgets to tell his mom and dad. When he gets scared he might pull his mom's hair or even a stranger's hair. This is not good because he might hurt the person and get into trouble.

Seth does not want to hurt people or get in trouble. Next time he gets scared at a restaurant he will say, "I'm scared." Then his mom and dad will know he wants to leave and eat outside or at home. He will feel better after he leaves.

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

All Behavior Serves A Purpose

Attention Seeking

Avoidance

Escape

If you battle against biology you will lose!



The Balance

- Behavior intervention is often unnecessary when etiology is considered
- Understanding “hard wiring” of students makes disciplining archaic
- Supporting students’ individual differences enables them to thrive, save face and take risks. Acting out, struggling for power and non compliance then become extinct

The Balance

Positive Support Plan

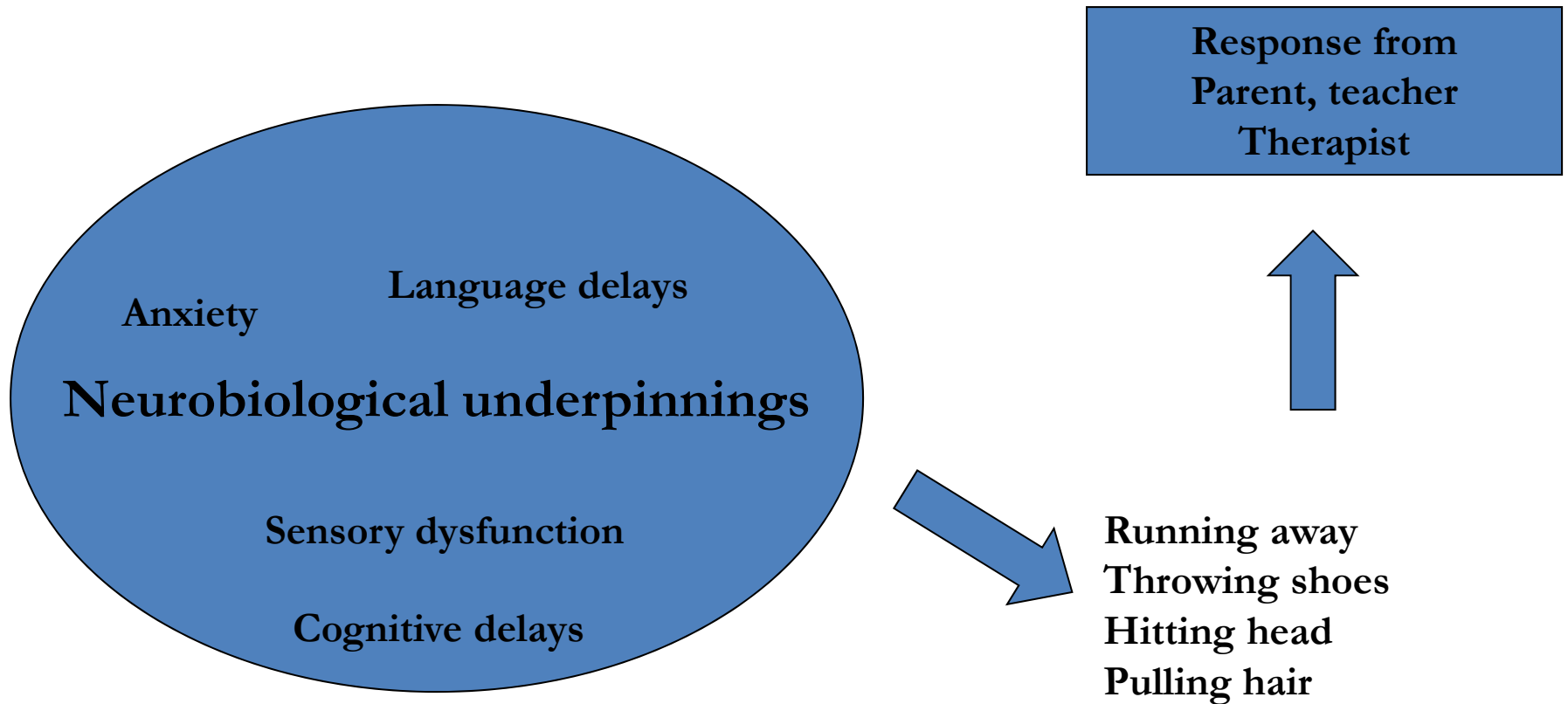
Behavior Intervention Plan



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

The behavioral cycle



Situation Specific Behaviors

- Provide as many external cues as possible
 - Changes of clothing, physical parameters
- When you give directions include the location
 - “That’s not okay at school.”
- Make the transitions clear
- Give an alternative location where the behavior is okay
- Be consistent
- Carefully balance how many situation specific behaviors are allowed
- Examples: wrestling mat, marshal arts uniform

FBA Summary

- Setting Event: What may be happening at home, on the way to school, before school?
- Predictor or Trigger: What sets him off?
- Presenting Problem: What does he do that is not appropriate?
- Consequence: What happens right after the inappropriate behavior?
- Maintaining Function: What does he want, why does he do what he does?

What is the function of this behavior?

- video

What is the function of this behavior?

- What is he avoiding?
- What is he trying to communicate?
- Why is he avoiding?
- What supports could be added to increase his engagement?

The importance of appropriate academic settings, curricula, and pedagogy

For issues within the classroom
examine

- Transitions
- Setting or physical environment
- Curriculum
- Pedagogy

A Successful Intervention

- How to go from this
(insert video)

A Successful Intervention

- To this.....
- Add video

How to Develop a Behavior Intervention Plan (BIP)

- Identify one specific behavior to target
- Determine when, where and how often it occurs
- Rule out any physiological causes
- Examine and modify structure when possible to decrease opportunities for behavior to occur
- Provide the child with additional resources for dealing with stressors associated with the behavior

How to Develop a Behavior Intervention Plan (BIP)

- Outline natural consequences for the behavior
- Be consistent
- Provide substitute behavior
- Reinforce positive behavior
- Start in a controlled setting
- Provide opportunities for both success and failure

Behavior Plan

- Reinforce positive behavior.
- Start in a controlled setting.
- Provide opportunities for both success and for failure.

Why is it important to use a consistent phrase?

- Language delay
- Consistency of language allows you to change location without disrupting routine
- You do not have to think about what you are going to say in the heat of the moment
- Keeps you from engaging in discussions and arguments
- It is calming to you
- Lets others know what you are doing
- Reminds you and your child of why they are in Time Out

Positive Behavior Support Plan

- Provide predictability of schedule as much as possible
- One to one intervention with small groups as appropriate
- Positive reinforcement
 - Tokens used to signal “on task” behavior and compliance to redirection
 - Points or tokens for delayed reinforcement (end of day)
 - Verbal or gestural reinforcement as needed to encourage success
- Limit transitions and provide support whenever necessary

Positive Behavior Support Plan

- Curricular adaptations will include high-strength academic materials. Staff will limit verbal input while pairing with visual support.
- Antecedents will be considered before intervention or consequences are applied
- Picture schedule with frequent breaks will be provided on a consistent basis.
- Use social stories to teach adaptive behavior

Total Melt Down

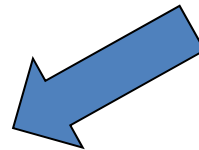
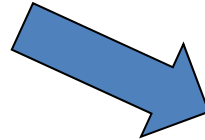
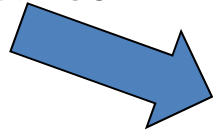
- Keep the child and others safe, including yourself
- Calming techniques and sensory integration activities will not work at this time
- Remove them from the cause or remove the cause from them if possible unless this is a “controlled burn”
- View it as a learning experience
 - What did you learn from this?
 - What did the child learn from this?
- Make a plan for the next time

Using Time Out

- Does not need to be a specific place
- Does need to be time away from your attention
- Needs to be linked to a specific act or behavior
- Needs to be used in a consistent manner
- Should be imposed immediately following the behavior
- Do not engage in repartee
- Do not address behaviors that occur on the way
- If he/she gets up put them back
- This is one component of a behavior plan
- Decide what you are going to say before you start

Behavioral Deterioration

Add pictures



Interrupt and Intervene

BEHAVIORAL CHAIN

- The pairing of a behavior with a response
- Expecting the same response whenever that behavior occurs
- Continuing or discontinuing a behavior based on the reaction to the behavior

Interrupting A Behavioral Chain

- Video

Interrupting A Behavioral Chain

GV attempts to intimidate by posturing aggressive behavior

GV is given a natural consequence for his behavior

STRATEGIES TO INTERRUPT A BEHAVIORAL CHAIN

MAKE AN ATTEMPT TO REDUCE ANY SENSORY INPUT

- Turn down the radio, stereo, TV and/or use white noise to filter out sound
- Reduce lighting, partial lighting, rheostat, close blinds, etc.
- Reduce verbal input immediately, wear sunglasses, avert your gaze or turn your back, give no gestural or facial feedback

STRATEGIES TO INTERRUPT A BEHAVIORAL CHAIN

INTRODUCE A DISTRACTOR

- Begin a novel, high interest video with muted sound or soft volume
- Play soft classical music, voice of sibling not present, or favorite TV show on audio tape (this can be done in the car when driving)
- Provide an album of funny pictures, costumes at Halloween, favorite vacations, etc.
- Present a hand held calculator and ask the individual with Fragile X to work out an addition problem $5+6+8+9+3$ etc. (Don't worry about the correct answer, remember this is just a distractor)

Understanding Your Own Scale



- Changing behavior is not easy
- The behavior will get worse before it gets better
- Understand the trade- offs
- You do not need to change everything now

Questions?