

No Longer Fragile

An education seminar on fragile X syndrome

Unit 1: Introduction to Fragile X

FraXgile



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Goals

- Learn about Fragile X syndrome
- Discuss how Fragile X affects your child
- Discuss possible strategies
- Record important information about the child
- Share information within your team

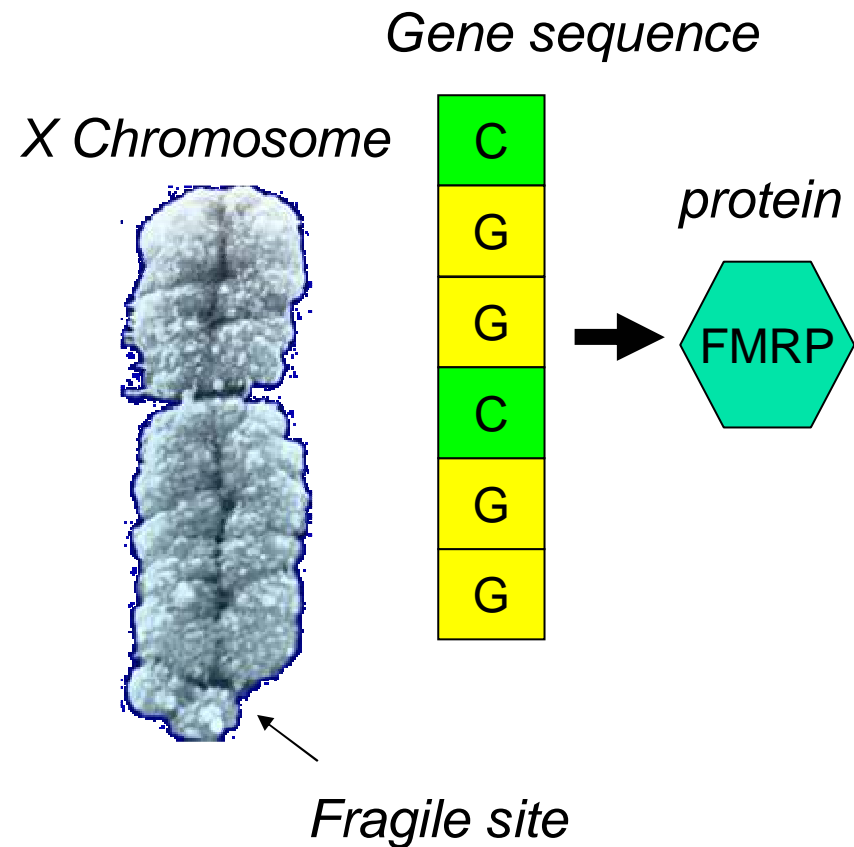


What is Fragile X Syndrome?

- Leading *inherited* cause of intellectual disability and developmental delay
- Affects 1 in 4000 individuals
- Leading known cause of autism
- Spectrum of impairment includes autism, anxiety, ADHD, mild to severe learning difficulties, language delay

What causes fragile X ?

- A gene sequence expands on the X chromosome
- Expansion causes the gene to turn off
- The gene is responsible for synthesis of a protein (FMRP) essential for normal brain function





FX Profile

- Anxiety / Hyperarousal
- Hyperactivity & attention difficulty
- Sensory sensitivities
- Cognitive profile
- Speech & language profile
- Difficulty with social interaction

FX children

- Good sense of humour
- Individuals
- Passionate interests
- Gentle hearts
- Interesting & challenging
- Children you can have a lot of fun with





Anxiety

The experts say:

- “Most prevalent and troubling characteristic of FX”
- “Decreases ability to think clearly and adapt to situations”
- “Must be directly managed as a part of life at school, work and home”

www.developmentalfx.org

Self Regulation



Hypoarousal



Arousal



Hyperarousal

- Our level of arousal affects our ability to perform any task
- Research has shown that fragile X children's normal arousal state is higher than their neurotypical pairs

Solution

- Recognise when our children are in a state of hyperarousal
- Recognise, remove or reduce triggers
- Regulation strategies



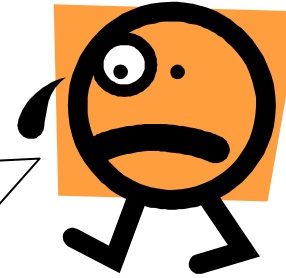


Anxiety: what does it look like?

- **Speech**
 - Perseveration
 - Increased rate & cluttered
 - Inappropriate words
- **Behaviour**
 - Hyperactive
 - Sensory seeking\avoiding
 - Gaze avoidance
 - Hand-biting
 - Hits out or runs away



I'm anxious when.....



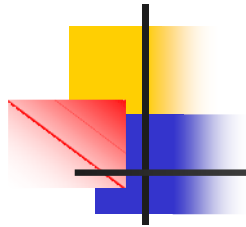
What does my child say?

- A repeated phrase
- I need the toilet
- I'm hungry
- I've got a sore tummy
- I'm not feeling very well

When & where?

- Move around room
- Lie down
- Tense body
- Bites fingers
- Hits out
- Runs away
- Cries
- Takes off clothes
- Covers ears
- Screams
- Covers face
- Averts eyes

What does my child's body do?



Triggers

- Negative emotional effect
- Sensory environment
- Change
- Transitions
- Behaviour chains
- Task
- Communication pressure
- Social pressure



Negative emotional affect

- Fragile X children may become upset by emotionally negative:
 - voice tone
 - body language
 - facial expression
 - other upset children
- They need to feel accepted by others



Sensory sensitivities

Individuals with fragile X often have an extreme over- or under-reaction to sensory input.

This can affect:

- Touch
- Sound
- Smell
- What or how they see
- Body awareness
- Need for movement



Seeking and avoiding

- Some sensory input will be experienced by the student as pleasant or calming
- Some sensory input will be experienced by the student as unpleasant or anxiety provoking
- Students will seek sensory input experienced as pleasant and avoid activities that are experienced as unpleasant



Touch

Your child may seek

- Control
- Messy play
- Specific textures
- Touch everything
- Use their hands to explore objects

Your child may avoid

- Soft touch
- Bare foot
- Hair cuts
- Wind
- Stuff on their hands or face
- Crowds



Sound

acute hearing, distracted by sound

Your child may seek

- Repetitive & predictable sound
- Train sounds
- White noise
- Small engine noise, egg beaters or dryers
- One song played over and over

Your child may avoid

- Sudden noise
- Uncontrollable noise
- Crowd noise
- A baby crying
- Alarms
- Driers

Visual

Your child may seek

- Visual movement
- Spinning objects
 - Egg beaters, spinning tops, wheels, fans
- Changes in color
- Light/shadow contrasts
- Certain colours
- Running water
 - Waterfalls
 - Fast flowing taps

Your child may avoid

- Visual clutter
- Bright light
- Eye contact



Vestibular

- The sense provided by receptors in the inner ear
- Provides information about head position
- Important for balance and movement





Vestibular

Your child may seek

- Fast movement
- Swings
- Flying fox rides,
- Spinning,
- Bouncing, running, dancing
- Car rides

Your child may avoid

- Putting their head back – in the pool, on a dentist chair or during hair washing
- Heights & stairs
- Elevators
- Car rides
- Climbing

Proprioception

- A sense provided by receptors in your joints and muscles
- Activated by heavy muscle work and deep pressure
- Provides information about where you are in space. Best sensory regulator





Proprioception

- Rough play
- Being squashed
- Squeeze between furniture
- Chew on objects, crunchy food apples, crackers or chewy food like steak.
- Playing drums
- Pushing or pulling
- Clenching their body
- Enjoy outside physical play – trampolines, jumping, kicking balls

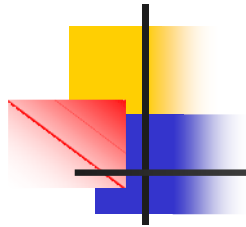


Sensory profile

- Describe the students sensory issues



How can we change the
environment to help?



Triggers

- Negative emotional affect
- Sensory environment
- Behaviour chain
- Transitions
- Change
- Task
- Communication Pressure
- Social Pressure



Behavioural Chains

- A behavioural chain is when a behaviour is part of a learnt routine or follows a visual or verbal cue.
- The best solution is to interrupt the event chain by distracting or changing the event sequence.

Strategies for transitions

- Provide a task for the child to complete
- Ask the child to help a buddy to complete a task
- Transition objects help calm
- Provide calming sensory input



Marcia Braden: www.marciabraden.com



Strategies for change

- Provide information about upcoming events
 - What it will look like
 - Who will be there
 - What will happen
 - Why
 - A part to look forward to



Strategies for change

- Use visual timetables
- Stories that explain what will happen
- Visit
- Act out the event sequence
- Look at photos, maps, pamphlets



Change and transitions

- What change or transitions cause anxiety?

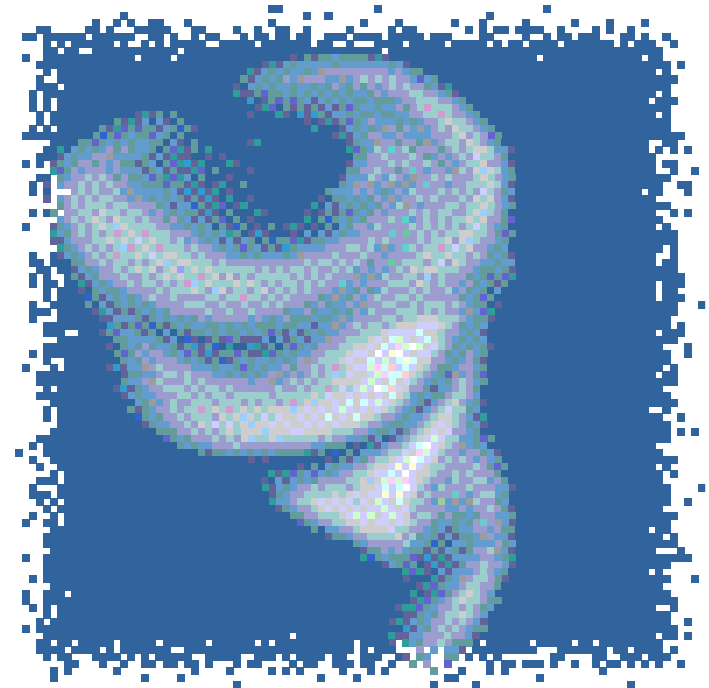
- What strategies can we put in place?

- Arriving at school
- Change to classroom arrangement
- Relief teachers
- Change in routines
- Getting out of the car
- Moving from one activity to the next
- Shopping
- Morning sequence



Emotional Regulation Plan

- Good days
 - Extend time on task
 - Expect more
 - Try something new
- Bad days
 - Reduced school day
 - Calm environment
 - Stick to routines
 - Lots of sensory breaks





Summary

- Anxiety needs to be directly managed
- Provide a calm, positive, learning environment
- Consider the child's sensory needs
- Reduce sound & visual distractions
- Provide regular sensory breaks
- Provide information about what will happen next
- Support change & transitions



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FRAGILE X TRUST (NZ)

*Supporting New Zealand
families living with
fragile X syndrome*