Stress & Anxiety Responses in Children with 22q11.2DS

Presentation Plan:

1. Provide an experiential activity to demonstrate some of the neurocognitive, behavioral and emotional challenges.

2. Provide information about some interventions that may ease these challenges and CABIL’s plans to evaluate their effectiveness.
Put yourself in their shoes

Notice how you feel and think as you experience these challenges…

- Visual-Spatial
- Attention and Working Memory
- Time
- Abstract Reasoning and Expressive Language
- Self-Regulation of
  - Sensory, Emotional and Behavioral processes
Preactivity: Participant’s Stress Symptoms Checklist

Take a personal inventory. Rate the intensity of your body sensations and your thought processes RIGHT NOW.

<table>
<thead>
<tr>
<th>Tense Muscles</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
<th>8</th>
<th>9</th>
<th>10</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bored</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td>5</td>
<td>6</td>
<td>7</td>
<td>8</td>
<td>9</td>
<td>10</td>
</tr>
</tbody>
</table>
When your rating is completed

- Put it aside for later comparison.
- Draw a ticket from the cup to discover what team you will join.
- Move to your team’s table.

<table>
<thead>
<tr>
<th>5 TEAMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>BAY LAUREL</td>
</tr>
<tr>
<td>BUCKEYE</td>
</tr>
<tr>
<td>ELDERBERRY</td>
</tr>
<tr>
<td>MADRONE</td>
</tr>
<tr>
<td>TANOAK</td>
</tr>
</tbody>
</table>
### Sneak A Peak Activity

<table>
<thead>
<tr>
<th>TEAMS</th>
<th>Activity Facilitators</th>
</tr>
</thead>
<tbody>
<tr>
<td>BAY LAUREL</td>
<td>Elliott</td>
</tr>
<tr>
<td>BUCKEYE</td>
<td>Freddy</td>
</tr>
<tr>
<td>ELDERBERRY</td>
<td>Ingrid</td>
</tr>
<tr>
<td>MADRONE</td>
<td>Kathy</td>
</tr>
<tr>
<td>TANOAK</td>
<td>Margie</td>
</tr>
</tbody>
</table>
Activity Facilitators

- Show the Director a target model for 15 seconds, then remove the model.
- Monitor the team and the rules while the Builder has 30 seconds to construct an exact replica using the Director’s oral instructions.
Example of Target Model
<table>
<thead>
<tr>
<th>Role</th>
<th>Image</th>
</tr>
</thead>
<tbody>
<tr>
<td>Director</td>
<td><img src="image" alt="Director" /></td>
</tr>
<tr>
<td>Builder</td>
<td><img src="image" alt="Builder" /></td>
</tr>
<tr>
<td>Guide</td>
<td><img src="image" alt="Guide" /></td>
</tr>
<tr>
<td>Observer/Recorder</td>
<td><img src="image" alt="Observer" /></td>
</tr>
<tr>
<td>Audience</td>
<td><img src="image" alt="Audience" /></td>
</tr>
</tbody>
</table>
Team Director

- Look at your team’s target model for 15-seCONDS.
  - (Only you and the activity facilitator may see it.)
- Try to memorize it before returning to your team.
- You have 30-seconds to tell the Builder how to make an exact replica.
Team Director- LIMITATIONS

- When you are instructing the Builder, you will wear a sleep mask so you may not see what the Builder is doing.
- You may not answer the Builder’s questions or otherwise converse with the Builder.
Builder

- Attempts to construct a replica by listening to and following the Director’s oral instructions.
- The Builder may tell the guide about how their body feels or their thoughts... BUT
Builder- LIMITATION

- The Builder may not “cross-talk” with the Director or Guide to ask questions about or comment on the replica.
- The Builder may not see the target construction.
Guide

- Orally describes the replica and its progress to the Director.
- Asks the Director to clarify instructions.
- Encourages the Builder to cope.
Guide: LIMITATION

- May not draw, touch or gesture to the construction materials.
- The Guide may not see the target model that the Activity Facilitator showed the Director.
Observer/ Recorder

- Watches and records the observable signs of stress in the Builder.
Observer/Recorder - LIMITATIONS

- The recorder is a silent observer.
- They may not give suggestions, ask questions, use gestures or otherwise help the Builder, Guide or the Director.
Audience

• Offers the Director, Builder and Coach support and analysis such as:
  – That was a good, clear instruction!
  – Slow down, you’re talking too fast.
  – You’re doing great, stay with it.
  – That’s confusing, name the shape you mean.
  – Don’t think about that Madrone Team, stay focused on our replica.
Audience- LIMITATIONS

- May not tell the Builder what to do.
- For example, they may not say
  - Build a rectangular base on the table with the two long yellow blocks and the two short red ones.
  - Now balance the triangular block on top of the two columns.
  - No, no, he means the other piece.
Role Assignment

- Each team identifies 4 volunteers to be a
  - Director,
  - Builder,
  - Guide and
  - Observer/Recorder.
- Others are Audience.
- Activity Facilitator distributes Role Tags.
After 30-seconds of building

- Activity Facilitator stops action and puts aside replica.
- All team members RATE their stress levels on the symptoms checklist.
- ROLES ARE ROTATED
  - Builder → Director
  - Director → Guide
  - Guide → Builder
- The activity starts again with the facilitator showing the new Director the target model.
### Everyone to your places

<table>
<thead>
<tr>
<th>Team</th>
<th>Activity Facilitator</th>
<th>Materials</th>
<th>Sensory Experience</th>
</tr>
</thead>
<tbody>
<tr>
<td>BAY LAUREL</td>
<td>Elliott</td>
<td>TRiO</td>
<td>Gloves</td>
</tr>
<tr>
<td>BUCKEYE</td>
<td>Freddy</td>
<td>Kid K’NeX</td>
<td>1812 Overture</td>
</tr>
<tr>
<td>ELMERBERRY</td>
<td>Ingrid</td>
<td>Flexeez</td>
<td>Pediatrics Lecture</td>
</tr>
<tr>
<td>MADRONE</td>
<td>Kathy</td>
<td>Flexeez</td>
<td>Popcorn</td>
</tr>
<tr>
<td>TANOKE</td>
<td>Margie</td>
<td>Legos</td>
<td>Mirror View</td>
</tr>
</tbody>
</table>
What is self-regulation?

• “An integrated set of abilities or skills that draw from both executive function and emotion regulation capacities, which are invoked in the service of accomplishing both proximal and distal goals.” (Buckner et al. 2009)

Self-Regulation is crucial to adaptive functioning
Executive Functions

• Executive functions include working memory, executive attention, inhibitory control, detecting novel stimuli, planning, set-shifting and decision making
Emotion Regulation

• “Emotion regulation can involve maintaining and enhancing emotional arousal as well as inhibiting or subduing it” (Thompson, 2004)
Heart Rate Variability

- Indexes
  - Attention/ Executive Function
  - Self-Regulation

- HRV also may be an independent variable that changes attention, executive function and self-regulation
Evidence Based Interventions

• Attention and Working Memory
  – Cogmed Working Memory Training

• Anxiety
  – Cognitive Behavior Therapy
    • (e.g., Coping Cat and Camp Cope-A-Lot)

• Heart Rate variability
  – Biofeedback
    • (e.g., Heart Math’s emWave Stress Relief System)
Cogmed Working Memory Training

• 25 Home-based, interactive on-line sessions
  – (30-45 minutes daily, 5-days/week for 5-weeks)
• 7-17 year olds who want to improve working memory
  – Also impacts attention, impulse control, complex reasoning and academic performance.
• Parent is Training Aide
• Weekly Check-in and Coaching
• Go to training demonstration
Cognitive Behavior Therapy for Anxiety

• 12 sessions, computer-assisted cognitive-behavioral therapy program.
  – Interactive, animated characters and videos instruct children how to identify bodily sensations associated with anxiety, learn how to change their internal, self-talk, and develop problem solving skills.
  – They also learn a systematic relaxation strategy and participate in therapist supported exposure tasks.

• Targets anxiety in youth 7-13 years old
• First 6-sessions self-guided, home-based
• Last 6-sessions therapist coached
• http://www.cope-a-lot.com/
Biofeedback

- Using principles of operant conditioning, individuals learn how to change physiological activity to improve health and performance.
- Physiological activity (heart function and respiration) is rapidly and accurately "fed back" to the user through precise instruments.
- Training occurs in clinic and in the individual’s natural environment.
- When skills are practiced regularly, changes can endure over time without continued use of an instrument.
Biofeedback

- Respiration: Abdominal & Thoracic
- Blood Volume Pulse/ Heart Rate Variability
- Skin Conductance
- Skin Temperature
- Surface EMG
- EEG
- Biofeedback Training and Equipment from Stens
  Biofeedback
Biofeedback

• Bradley et al. (2010) HeartMath
  – Heart Rate Variability
  – Coherence
Portable Home Training

- Heart Rate Variability and Coherence
  - emWave Training
    - [http://www.heartmathstore.com/item/1322/emwave-coherence-system](http://www.heartmathstore.com/item/1322/emwave-coherence-system)
  - StressEraser

- Paced Breathing Blood Pressure
  - RESPeRATE
Technology Facilitates Remote Delivery of Interventions

• CABIL Translational Research Impact Survey
  – Parents report challenges finding practitioners who are familiar with and adequately trained to offer evidence based interventions
Are Evidence-Based Interventions Useful?

- Cogmed
  - Generally not recommended for individuals with Intellectual Disability, Anxiety, Oppositional Defiant Disorder
  - Moss (2010)
Are Evidence-Based Interventions Useful?

- CBT
  - Ability to reason abstractly, understand and use language may impact outcome
Are Evidence-Based Interventions Useful?

- Biofeedback of Heart Rate Variability and Coherence
  - Does not require reasoning or language
  - Behavior changes is based on operant conditioning
Research Questions

• Do these programs differentially benefit children with 22q11.2 DS?
  – Do programs that train more proximal, micro-level skills (e.g., HRV biofeedback and Cogmed) benefit children with 22q11.2DS more broadly than programs that train macro-level metacognitive skills (e.g., Camp Cope-A-Lot).

• No matter the intervention, children whose heart rate variability and the synchrony between heart rhythm and respiration improve most, will show the largest gains in adaptive functioning, attention, working memory and will experience fewer anxiety symptoms.
Research Questions

- Do children with different cognitive levels benefit more from one intervention than the other?
Recruitment

- Nine children (6 participants, 3 back-ups)
  - Budget limits the number we can serve
- 7-14 years with 22q11.2 DS residing in the Northern California/Western Nevada and within a 2.5-hours drive from Sacramento.
  - Age
    - We will attempt to match all 6 participants on age, within a band of +/- 30-months
  - Two levels of function
    - average IQ or higher and borderline IQ or lower
## Research Design

<table>
<thead>
<tr>
<th>Epoch 1</th>
<th>Pre-intervention Assessment (Week 1-4)</th>
<th>Intervention (Week 5-15)</th>
<th>Post-intervention Assessment (Week 16)</th>
<th>6-month Follow-Up (week 40)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child 1</td>
<td>BFDBK</td>
<td>Child 1</td>
<td>Child 4</td>
<td></td>
</tr>
<tr>
<td>Child 2</td>
<td>COGMED</td>
<td>Child 2</td>
<td>Child 5</td>
<td></td>
</tr>
<tr>
<td>Child 3</td>
<td>CCAL</td>
<td>Child 3</td>
<td>Child 6</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Epoch 2</th>
<th>Pre-intervention Assessment (Week 12-15)</th>
<th>Intervention (Week 16-26)</th>
<th>Post-intervention Assessment (Week 27)</th>
<th>6-month Follow-Up (week 51)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child 4</td>
<td>BFDK</td>
<td>Child 4</td>
<td>Child 4</td>
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</tr>
<tr>
<td>Child 5</td>
<td>COGMED</td>
<td>Child 5</td>
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<td></td>
</tr>
<tr>
<td>Child 6</td>
<td>CCAL</td>
<td>Child 6</td>
<td>Child 6</td>
<td></td>
</tr>
</tbody>
</table>

Note: BFDBK = Biofeedback Training, COGMED = Cogmed Working Memory Training, CCAL = Camp Cope-A-Lot Computer Assisted Cognitive Behavioral Therapy
Pre-, Post- and Follow-up Measures

- Functional Skills
- Attention
- Anxiety
- Diagnostic Classification
- Psychophysiology