Team Members

Tony J. Simon, Ph.D. – Director, Cognitive Neuroscientist
Kathy Angkustsiri, M.D. – Developmental Behavioral Pediatrician
Ingrid Leckliter, Ph.D. – Psychologist
Khyati Brahmbhatt, M.D. – Child & Adolescent Psychiatrist
Elliott A. Beaton, Ph.D. – Postdoctoral Scholar
Michelle Deng, Ph.D. – Postdoctoral Scholar
Heather Shapiro B.S. – Graduate Student
Joshua Cruz, B.S. – Research Assistant
Not pictured – Janice Enriquez, Ph.D. – Psychologist; Margie Cabaral, B.S. – Research Assistant

Staffing

- Who attends?
  - Conference with clinicians and research team

- What do we discuss?
  - Assessment results and overall impressions
    - Individualized for each child
    - In the context of:
      - Child’s environment (family, community, school)
      - Other children with 22q11.2DS
  - Intervention recommendations
    - We all learn from each other to everyone’s benefit
Illustrative Case Composites*

“Coper”
Despite intellectual disability and poor academic skills this child is coping well. She is integrated into her community, is well-liked and has a positive sense of self-esteem. She also has good rote learning skills that support the development of basic academics (e.g., word reading, spelling).

*Identifying details are changed and data are composites, not an actual case.

“Struggler”
Despite average intelligence and academic skills, this child is anxious, has low self-esteem, is viewed by others as inattentive, disorganized, oppositional and unmotivated. She engages in risky behaviors.

*Identifying details are changed and data are composites, not an actual case.
A “Coper”- Case details

- 9-year, 9-months old female in 3rd grade (repeated second)
- Background:
  - Ventricular Septal defect (VSD)
    - No surgery needed
  - Submucous Cleft Palate
  - Thymic hypoplasia
    - Frequent infections when younger

A “Coper” (cont’d)

- Diagnosed with mild intellectual disability
  - INTELLECTUAL WISC-IV
    - FSIQ= 63, VCI= 65, PRI= 61, WMI= 83, PSI= 77
  - ADAPTIVE ABAS-II
    - GAC= 64, Conceptual= 69, Social= 84, Practical= 56
- Grade-based academic achievement WIAT-II
  - READING Composite= 80
    - Word reading= 90, Pseudoword reading= 82, Reading Comprehension= 74
  - MATH Composite= 58,
    - Numerical operations= 63, Math reasoning= 64
  - WRITTEN LANGUAGE Composite= 83
    - Spelling= 98, Written expression= 72
- Language CELF-4
  - Core LANGUAGE Composite= 58
    - Recalling sentences and expressive vocabulary were relative personal strengths (both were scaled scores= 5)
A “Coper” (cont’d)

- ANXIETY SCAS- child & -parent reports (at risk*= 60-69, clinically significant** > 70)

<table>
<thead>
<tr>
<th>Scale</th>
<th>Child</th>
<th>Parent</th>
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</thead>
<tbody>
<tr>
<td>Panic / Agoraphobia</td>
<td>56</td>
<td>45</td>
</tr>
<tr>
<td>Separation Anxiety</td>
<td>53</td>
<td>47</td>
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<tr>
<td>Physical Injury</td>
<td>57</td>
<td>50</td>
</tr>
<tr>
<td>Social Phobia</td>
<td>38</td>
<td>40</td>
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<tr>
<td>Obsessive / Compulsive</td>
<td>57</td>
<td>45</td>
</tr>
<tr>
<td>Generalized Anxiety</td>
<td>53</td>
<td>47</td>
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Her family modeled and the child used language to restructure thinking and associated emotional reactions.

- The coping philosophy stemmed from the family’s cultural background in Confucianism.
- Feelings always change. So when you feel distressed, just do something else (while you wait for the feeling to change) and then notice that you don’t feel the same way anymore.
A “Coper” (cont’d)

- Their community was small, people knew and supported each other.
  - This is the philosophy behind the inclusion model of special education.
  - The family had friends in the community who were educators at the child’s school.
    - The child’s mother volunteered at the school (but in classes other than her daughter’s class).
  - Many of the child’s classmates had been the same since kindergarten.
    - They knew she needed help and valued her contribution.

- The child’s involvement in the performance arts provided a sense of belonging.
- She felt valued for making a personal contribution.
  - She performed traditional dances from her cultural background.
    - All looked forward to watching her perform.
    - She found personal esteem and sense of purpose in her strengths.
A “Coper” (cont’d)

- The family scaffolded their daughter’s attention to personal hygiene and looking attractive.
  - Although her possessions and routines were haphazard (when left to her own devices), this child’s family developed a visual schedule to help cue her into self-care routines and to assure appropriate personal hygiene.
  - They helped her select clothing similar to her peers (but within their limited budget) and consistent with her cultural identity.
    - Sometimes her appearance was pseudo-mature so the CABIL team discussed the risks this might engender.

A “Struggler”- Case details

- 14-year, 3-month old female
- Background:
  - Tetralogy of Fallot, repaired
  - Pharyngeal flap
  - Early intervention until age 3, now in mainstream 9th grade
  - Easily fatigued, complains often of feeling tired
A “Struggler” (cont’d)

Behaviors:
- “Social Butterfly” especially on-line with friends from former school
  - Recently moved to new school and is worried about making friends there
- Sullen and withdrawn at home
- Meltdowns during homework sessions with mother
  - Overwhelmed easily, works for 5 hours each night
- Anxiety
  - Will not use public bathrooms
  - Problems sleeping
    - worried about school the next day
    - worried about former classmate who was raped
- Repetitive behaviors
  - Cleaning
  - Sucks and picks the skin around her fingernails,
  - Rocks her body

Other:
- Attractive teenager; well-developed
- Dresses older than her chronological age
- Is making new friends
  - Has attended sleep-overs
- Eager to please, but also naïve
  - Follows others without any awareness of consequences
  - Sneaks out of the house
A “Struggler” (cont’d)

- Family history:
  - Alcohol abuse (Grandfather and aunt)
  - Mental illness (Uncles)

- Social history:
  - Parents divorced
    - Visits her father during summer recess and alternate major holidays
  - Has unstable relationships with her mother’s partners

- Average Intellectual Ability
  - INTELLUCTUAL WISC-IV
    - FSIQ= 91, VCI= 93, PRI= 106, WMI= 88, PSI= 83
  - ADAPTIVE_ABAS-II
    - GAC= 66, Conceptual= 67, Social= 84, Practical= 62

- Grade-based academic achievement WIAT-II
  - READING Composite= 97
    - Word reading= 105, Pseudoword reading= 104, Reading Comprehension= 91
  - MATH Composite= 87
    - Numerical operations= 95, Math reasoning= 83

- Language CELF-4
  - Core LANGUAGE Composite= 94
A “Struggler” (cont’d)

- ANXIETY SCAS- child & -parent reports (at risk*= 60-69, clinically significant** ≥ 70)

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<table>
<thead>
<tr>
<th>Subscale</th>
<th>Within normal limits</th>
<th>Probable Difference</th>
<th>Definite Difference</th>
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<tbody>
<tr>
<td>Tactile sensitivity</td>
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</tr>
<tr>
<td>Taste/ smell sensitivity</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Movement sensitivity</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Under-responsive/ seeks sensation</td>
<td>X</td>
<td></td>
<td></td>
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<tr>
<td>Auditory filtering</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Low energy/ weak</td>
<td>X</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Visual/ auditory sensitivity</td>
<td>X</td>
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A “Struggler”- Discussion

- No supportive services because academic scores on standardized tests are within normal limits and commensurate with her ability.
  - But clearly she has problems with everyday skills.
  - Anxiety plays a role in everyday impairment.
  - Excessive fatigue (anxiety and poor sleep, mental fatigue).
- Encouraged school to consider eligibility for special education services under the categories
  - Other Health Impaired (OHI) and/or
  - Significant Emotional Disorder (SED)

Mismatch between expectations and ability

- Child
  - Low self esteem
    - Poor grades, homework requires excessive time
- Parent
  - Mother notices differences in behaviors with mother vs. others (moodier, oppositional)
- Teacher/school
  - Performs poorly day to day (e.g., does not finish class work or turn in homework)
  - Anxiety/avoidance interpreted as lazy, unmotivated, distracted and disorganized
Other Issues

- Sensory integration behaviors misinterpreted
  - Rocking
    - Teachers interpreted as masturbating
  - Repetitive behaviors
    - Cleaning - Maladaptive anxiety coping
  - Friendly poke or elbow to the side
    - Does not accurately judge her own force/ strength
    - Out of context, may be considered aggressive or may provoke another teenager

EvStress/Anxiety Intervention

Evidence Based Behavioral Approaches
  - Cognitive Behavioral Therapy (CBT)
    - high linguistic/conceptual demands
  - Biofeedback, Meditation, Exercise …
  - Work with (any) good clinician

Pharmacological Approaches
  - SSRI - Selective Serotonin Reuptake Inhibitors
  - Stimulants - ADHD medications
    - reasonable concerns but safe & effective
  - Work with pediatrician/psychiatrist