Anxiety symptoms in children with chromosome 22q11.2 deletion syndrome and its effect on functioning

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Outline

- Background
- Anxiety symptoms in 22q11.2DS
  - Under identified despite known risk
  - Affect everyday life skills
  - Case example
Developmental-Behavioral Concerns in 22q11.2DS

- Developmental Delays
  - Motor delays
  - Speech and language delays
  - Learning disabilities
    - FSIQ low-average to borderline range
    - Sometimes misleading b/c usually verbal IQ better than performance IQ
    - Difficulties with planning, math, and visuospatial tasks
  - Social immaturity

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Developmental-Behavioral Concerns

- Behavioral concerns
  - Attention Deficit/Hyperactivity Disorder (ADHD) 30-50%
    - Easily distracted
    - Impulsive
  - Screening for psychiatric problems
    - Anxiety (40-60%), Depression, Bipolar, Schizophrenia/schizoaffective
  - Autism & Autistic Spectrum Disorders
    - Reported in 20-50%, mostly PDD-NOS
Anxiety Disorders in VCFS/22q11.2DS: 40-60%

- Specific phobias (up to 60%)
- Generalized Anxiety Disorder (33%)
- Separation Anxiety Disorder (15%)
- Obsessive Compulsive Disorder
- Social Anxiety

Anxiety can manifest as other behaviors
- Somatization, Inattention, Avoidance, etc.
Anxiety

- Anxiety symptoms are under-identified
- Anxiety affects everyday living skills
  - Interventions available
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- 15 of 79 (19%) had a prior diagnosis of anxiety disorder
  - 6 (40%) meds
  - 7 (47%) therapy

- 46 of 79 (58%) currently have significant anxiety symptoms

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Adaptive function

- Measure of self-sufficiency and the ability to manage everyday needs in communication, self-care, etc.
  - According to developmental level
    - Self-care skills
    - Getting dressed
    - Asking for help
    - Riding a bus
    - Ordering food in a restaurant
- Predicts later functioning in children with disabilities (Harrison 1987)
In most populations, adaptive function is correlated with IQ

- **Typical children**
  - VABS and KABC (Sparrow, Balla, Cicchetti, 1984)

- **Developmental Delay**
  - Intellectual Disability
    - Moderate correlation SB-IV and VABS (Dacey, Nelson, and Stoeckel 1999)
  - Autism
    - SB-IV and VABS (Carpenieri and Morgan 1996)
  - Down Syndrome (Loveland & Kelley 1991)

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- 78 children
- Ages 7-14 years
  - Mean age 10.5 years
  - 42 (54%) male
- Measures
  - WISC-4
  - Behavior Assessment System for Children-2\textsuperscript{nd} edition (BASC-2)
  - Adaptive Behavior Assessment System-2\textsuperscript{nd} edition (ABAS-2)
  - Spence Children’s Anxiety Scales
No relationship between FSIQ and adaptive function (BASC-2)

N=78, r=0.03; p=0.74

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Lower anxiety, higher adaptive function (BASC-2)

N=78, r=-0.27; p=0.017
Lower anxiety scores are related to better adaptive functioning
Panic Agoraphobia

N=34, r=-0.38, p=0.028 (parent)
Lower anxiety scores are related to better adaptive functioning.

Obsessive Compulsive

N=34, r=-0.47, p=0.005 (parent)
Separation Anxiety, Physical Injury

N=34, r=-0.32, p=0.07 (parent)  
N=34, r=-0.34, p=0.05 (parent)
Other anxiety measures no relationship with adaptive Social Phobia, Generalized Anxiety

N=34, r=0.006, p=0.97 (parent)  
N=34, r=0.05, p=0.78 (parent)

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Cognitive reserve and use of resources

- Resiliency against brain damage
  - Prevalent in Alzheimer’s research
  - Tasks are processed more efficiently

- Chronic stress, possibly in response to anxiety, has deleterious effects on the brain

- Dealing with anxiety uses up cognitive resources, making it more difficult to perform usual tasks
“Sally”

- This case illustrates the impact of emotional dysregulation on functional living, despite the presence of mostly average cognitive and achievement skills.
“Sally”

- **Background:** 10y11m ♀, h/o TOF, pharyngeal flap, E.I. until 3 years, now in 5th grade mainstream class

- **Behaviors**
  - **School** - no behavior problems, likes school
    - Worried about the start of every school year
    - Shakes and poor eye contact in front of class when presenting
    - Teacher concerns inattentive/hyperactive symptoms?
  - **Home** - repetitive behaviors (cleaning)
    - Easily upset, overreacts, and dysregulated (sounds, etc.)
      - Oppositional, tantrums
      - Sibling conflict
    - Perseverates
    - Scared of the dark (sleeps with lights on)
    - Difficulty falling asleep (“brain is thinking”); nightmares
    - Worries about parents being kidnapped

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“Sally”

RESEARCH EVALUATION

- Observations: shy, quiet at first, warmed up and was chatty worked hard, although seemed fatigued at times
- WISC-IV Index scores range: 80 (PSI), others in the 90’s
- WIAT-II Reading Composite= 84; Math Composite= 86, Written Language 97
- SCQ: 2
- Short Sensory Profile: differences in 4 of 7 domains
- ABAS-II: GAC 74, Conceptual=85, Social=75, Practical=74
  - Never starts conversations on topics of interest to others
  - Never initiates games liked by friends or family
  - Never organizes a fun activity without help
  - Never decides alone to join an organized group
  - Never controls anger when another person breaks the rules
  - Never keeps working hard on tasks without becoming discouraged
“Sally”

- Spence-
  - child: panic, separation anxiety, physical injury, obsessive compulsive
  - parent: separation anxiety, physical injury (90), generalized anxiety (81)
“Sally”

- If not managed, Anxiety → “inattentive”/noncompliant, oppositional/defiant behaviors, catastrophic reactions, bizarre coping responses
- Management involves caring for the individual as well as her system (e.g., how educators and parents interpret her behaviors).
“Sally”

- Management recommendations
  - CBT (e.g., Coping Cat) with family component
  - Consider SSRI
  - Occupational Therapy with Sensory Integration emphasis
  - Meet with school for Learning Disorder intervention
  - “Inattention” may be secondary to anxiety in this case

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Cognitive Behavioral Therapy

- Teach coping skills with self-instruction and behavioral techniques
  - Imagery, relaxation, modeling, role playing
  - Recognize anxious feelings and somatic reactions
  - Clarify cognition (unrealistic expectations, etc.)
  - Create a plan of action

- [http://www.abct.org/dHome/](http://www.abct.org/dHome/)
- Coping Cat, Camp Cope-A-Lot
  - Manualized, evidenced-based CBT (Kendall 1994)
  - Shows animated characters who think aloud and solve problems using a systematic structure
CAMP COPE-A-LOT

Problem: How to get flag down from tree

- Climb the tree
- Throw rocks at the flag
- Throw a rope to pull branch down
- Get counselor

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Resources

- Donna Cutler-Landsman, MS “Educating Children with VCFS”
- Carol Gray “Social Stories”
  - [http://thegraycenter.org/](http://thegraycenter.org/)
- Michelle Garcia Winner
- CABIL video
UC Davis M.I.N.D. Institute

Cognitive Analysis and Brain Imaging Laboratory (CABIL)

- BREAKING NEWS
- CABIL Team Members
- Publications by CABIL Team
- Research Projects at CABIL
- What Will My Child Experience at CABIL?
- Family Meetings
- Resources
- Directions
- Media Coverage of CABIL
- Research Tools

The M.I.N.D. Institute's Cognitive Analysis and Brain Imaging Laboratory (CABIL, pronounced “cable”) is directed by Dr. Tony J. Simon and funded by the National Institutes of Health. CABIL's mission is to investigate, explain, and eventually treat the impairments in cognitive function experienced by children with neurodevelopmental disorders.

The cognitive analysis part of our research involves developing theory-driven experiments that are presented to children as computer games. These experiments test the functioning of specific brain circuits under different conditions and predict characteristic patterns of performance depending on how well the system is functioning. The brain imaging part of our research is achieved using safe, radiation-free neuroimaging methods to characterize the changes in brain development that affect the neural structure, connectivity and function of such children. Knowing how the brains of children with neurodevelopmental disorders differs from those of their typically developing peers helps us to generate possible explanations for the impairments in cognitive function.

Click here to play a video to learn more about CABIL (requires Adobe Flash)
Thank You!

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