Cognitive Features of XY Variations

• Lowering of IQ compared to general population
  ▪ XXY/KS IQ 89-102
  ▪ XYY IQ 75-107
  ▪ XXYY IQ 70-80
  ▪ XXX IQ 85-90

• At risk for...
  ▪ Academic problems and learning disabilities
  ▪ ADHD
  ▪ Autism Spectrum Disorder
  ▪ Social, emotional, and behavioral difficulties
  ▪ Adaptive functioning deficits

• Executive functioning deficits
Cognitive Features of XY Variations

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• Executive functioning deficits
What are executive functioning skills?

• Processes involved purposeful, goal-directed behavior
  ▪ Planning and sequencing of complex behaviors
  ▪ Capacity for grasping the gist of a complex situation
  ▪ Ability to pay attention to several components at once
  ▪ Resisting distraction
  ▪ Inhibiting inappropriate responses
  ▪ Sustaining behavior for prolonged periods of time

• Can be considered the “command center” or “conductor” of all cognitive skills
Behavior Regulation

- *Inhibit*: stop an action or not react to impulse

- *Self-monitor*: aware of effect your behavior is having on others and change your behavior accordingly
Emotion Regulation

- **Shift**: move from one task or situation to another; transitions; flexible problem-solving

- *Emotional Control*: regulate emotional response
Cognitive Regulation

- **Initiate**: begin task; independently generate ideas
- **Working Memory**: hold information actively in mind
- **Plan/Organize**: anticipate future events and develop steps to attain goals; establish and maintain order
- **Organization of materials**: keeping belongings organized
- **Task monitor**: attend to your own performance on tasks (for example, checking for errors)
What do EF problems look like?

• The “walking tornado”

• Forgetting to turn in homework, even if it’s completed

• Difficulty completing long-term school projects or following complex directions

• Losing track of time; not using time efficiently

• Many times, kids with EF problems are viewed as “lazy” or “not trying” because they cannot complete basic tasks
EF SKILLS IN CHILDREN WITH XY VARIATIONS
Executive Functioning Skills in Boys with XXY and XXYY
Executive Functioning Skills in Boys with XXY and XXYY
Relationship Between EF and Adaptive Skills in Boys with XXY

• How do executive functioning deficits contribute to problems with daily living skills?

• Parents completed questionnaires looking at both EF and adaptive skills

• Cognitive regulation related to communication skills
  ▪ Initiating a conversation
  ▪ Organizing what to say
  ▪ Keeping “in mind” what is being said in the conversation

• Behavior regulation related to daily living skills and social skills
  ▪ Inhibiting behavior to get along with others (ex, not hitting)
  ▪ Shifting/changing behavior to meet demands of situation
Relationship Between EF and Behavior Problems in XXY and XXX

• How do EF problems contribute to behavior problems?

• Neuropsychological testing to assess EF, as well as parent questionnaires assessing EF and behavior problems

• Both groups showed deficits in inhibition, flexibility, sustained attention, and working memory

• Deficits in inhibition related to aggressive behavior and rule-breaking behavior
Development of Executive Functions

• Development not linear but occurs in “spurts”

• Frontal brain development continues into 20s
  ▪ EF development is protracted
  ▪ Major changes in brain structure between 13 and 19 years

• EF domains have different developmental trajectories
  ▪ Babies and young children have basic inhibitory control and attentional control
  ▪ Cognitive regulation skills develop later, starting around 7-8 years
5-year-old brain | Preteen brain | Teen brain | 20-year-old brain

Dorsal lateral prefrontal cortex ("executive functions")

Front

Top view

Back

Red/yellow: Parts of brain less fully mature

Blue/purple: Parts of brain more fully mature

Sources: National Institute of Mental Health; Paul Thompson, Ph.D., UCLA Laboratory of Neuro Imaging

Thomas McKay | The Denver Post
Environmental “Development”

• Increasing EF demands in everyday tasks

• As child becomes older, more demands for...
  ▪ Multi-step tasks/learning
  ▪ Novelty (lack of experience base) vs automaticity (familiar)
  ▪ High vs low stress
  ▪ Future vs present orientation

• Teachers/parents have expectations that specific skills will be “on-line” by certain developmental periods
What happens when development is not typical?

• Child may not meet environmental expectations because of EF deficits

• “Mismatch” between child’s abilities and expectation of the environment

• Greatest difficulty seen when there are dramatic increases for...
  ▪ Independent work
  ▪ Rapid and efficient processing of information
# Academic Tasks and Potential Stress Points

<table>
<thead>
<tr>
<th>Time Point</th>
<th>Task</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kindergarten</td>
<td>Learning to learn</td>
</tr>
<tr>
<td>1&lt;sup&gt;st&lt;/sup&gt; grade</td>
<td>Learning to read</td>
</tr>
<tr>
<td>4&lt;sup&gt;th&lt;/sup&gt; grade</td>
<td>Reading to learn</td>
</tr>
<tr>
<td>Middle School</td>
<td>Learning to organize</td>
</tr>
<tr>
<td>High School</td>
<td>Learning to read, learn, organize on own</td>
</tr>
<tr>
<td>College</td>
<td>Doing it on own</td>
</tr>
</tbody>
</table>
Basic Principles of EF Interventions

• Skills don’t come intuitively, so we must explicitly teach them

• Teach a process of systematic thinking

• Teach child how to learn and approach tasks
- Consistency across settings
- Explicitly teach skills
- Teach in structured, systematic way

Teach and use key scripts and words

Teach by doing

Use visual supports and routines

Make it fun!

- Be an active coach
- Everyday people, everyday activities
- Model strategies
- Support-fade-generalization

- Checklists
- Cues
- White boards

- Humor, rewards
- Collaborate with child

From Kenworthy et al., *Solving Executive Functioning Challenges*
Consider the Demands of the Task or Situation

• Task analysis to help figure out where the breakdown occurs

• “Task” could be academic, around house, social, or behavioral
  ▪ Remember, seemingly simple tasks can have multiple EF demands making it difficult for children with EF deficits (for example, showering)

• What are the EF demands of this task?
  ▪ Multiple steps or multi-tasking?
  ▪ Require planning ahead of time?
  ▪ Require child to break a larger task into smaller chunks?
  ▪ Change behavior in response to feedback from others?
  ▪ Require child to change behavior or change situations?
Two Levels of Intervention

• Intervention at the level of the environment
  ▪ Changes adults and teachers can implement
    • Change environment (reduce distractions)
    • Change nature of task (make it more structured)
    • Change way interact with child (anticipate problems)
  ▪ Provide modeling and scaffolds that are eventually reduced as child gains skills

• Intervention at the level of the child
  ▪ Explicit instruction in EF strategies
  ▪ Giving a “tool kit” of strategies the child can apply to different situations
Working Memory

• Decrease working memory “load”
  ▪ Keep instructions clear and concise
  ▪ Give instructions one step at a time; written and oral format-
    provide a “hard copy”
  ▪ Teach information in smaller chunks
  ▪ Repeat instructions as needed

• Encourage child to repeat/paraphrase what was said and
  write down steps while completing a task (like a math
  problem)

• Use of memory aids- notebooks, calendars, electronic
  devices

• Teach active listening and active reading strategies
  ▪ Stop what doing; focus attention; ask questions; relate
    information to question
  ▪ Write down themes, characters, etc., while reading
Planning

• Planning problems often a result of not knowing what step to take next in a task

• Break long-term projects into clearly defined subtasks

• Provide template/ list of steps to be followed
  ▪ Younger children use photos or symbols
  ▪ “Cookbook” for common tasks

• Set aside “planning time” each day for older students to plan assignments

• Have child verbalize plan before starting a task
  ▪ Places time between thought and action, decreasing impulsivity

• Teach specific routines to identify goals and plan steps to attain goal- at first, work through the planning process together
  ▪ Early stages, questions need to prompt each step
    ▪ What do you have to do first?
    ▪ What do you have to do next?
  ▪ With success, prompts become more general
    ▪ “Let’s make a list of all the things you have to do to complete this project”
    ▪ Over time, child internalizes ability to make a plan more independently
# Desk Cleaning Checklist

Step 1: Gather Necessary Materials

<table>
<thead>
<tr>
<th>Materials Needed</th>
<th>Check all that apply</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wastebasket</td>
<td></td>
</tr>
<tr>
<td>Empty file folders</td>
<td></td>
</tr>
<tr>
<td>Three-ring binders</td>
<td></td>
</tr>
<tr>
<td>Paper clips</td>
<td></td>
</tr>
<tr>
<td>Stapler</td>
<td></td>
</tr>
<tr>
<td>Manila envelopes</td>
<td></td>
</tr>
</tbody>
</table>

Dawson & Guare (2004)
## Desk Cleaning Checklist

**Step 2: Follow Desk Cleaning Procedure**

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Check when done</th>
</tr>
</thead>
<tbody>
<tr>
<td>Empty out desk</td>
<td></td>
</tr>
<tr>
<td>Sort everything into two piles: <em>Save/ Don’t Save</em></td>
<td></td>
</tr>
<tr>
<td>Throw <em>Don’t Save</em> pile into wastebasket or recycle bin</td>
<td></td>
</tr>
<tr>
<td>Sort <em>Save</em> into two piles</td>
<td></td>
</tr>
<tr>
<td>1. School stuff (books, assignments, pens, etc)</td>
<td></td>
</tr>
<tr>
<td>2. Home stuff (notices for parents, uneaten snacks, etc)</td>
<td></td>
</tr>
<tr>
<td>Put “home stuff” into backpack (uses folders if needed)</td>
<td></td>
</tr>
<tr>
<td>Sort “school stuff”, one pile for each subject</td>
<td></td>
</tr>
<tr>
<td>Organize each subject following teacher instructions</td>
<td></td>
</tr>
<tr>
<td>Put school materials neatly back in desk</td>
<td></td>
</tr>
</tbody>
</table>
## Mission Possible

**Clean Room Checklist**

<table>
<thead>
<tr>
<th>Steps to a clean room</th>
<th>Check it off!</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Pick up clothes</td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td></td>
</tr>
<tr>
<td>5.</td>
<td></td>
</tr>
<tr>
<td>6.</td>
<td></td>
</tr>
<tr>
<td>7.</td>
<td></td>
</tr>
<tr>
<td>8.</td>
<td></td>
</tr>
<tr>
<td>9. Get my reward!</td>
<td></td>
</tr>
</tbody>
</table>

**Figure 4.5.** Mission Possible: Clean Room Checklist.

*Solving Executive Function Challenges: Simple Ways to Get Kids with Autism Unstuck and on Target,* by Lauren Kenworthy, Laura Gutermuth Anthony, Katie C. Alexander, Monica Adler Werner, Lynn Cannon, & Lisa Greenman. Copyright © 2014 by Paul H. Brookes Publishing Co., Inc. All rights reserved.
From Kenworthy et al., *Solving Executive Functioning Challenges*
Behavior Regulation

• Environmental modifications
  ▪ Sitting near the teacher, away from distractions
  ▪ Teacher check-ins
  ▪ Regular breaks (running errands for the teacher)
  ▪ Emphasize quality of work rather than quantity
    • Place check box on top of worksheet to show they checked for errors

• Behavior regulation strategies
  ▪ Teach response delay techniques
    • Counting to 10 before acting
    • Leaving the situation
  ▪ Stop-Think-Talk-Do
    • “Stop” behavior before acting impulsively
    • “Think” about cause-and-effect relationship of intended behavior
    • “Say” or verbalize to self or others what they will do
    • “Do” the chosen behavior

• If diagnosed with ADHD, medication may be appropriate
Social Skills EF Interventions

• EF skills help us navigate the social world
  ▪ What EF deficits are contributing to social difficulties?

• Model and discuss your own social concerns/worries and your solutions for dealing with them

• Provide additional structure to social situation

• Provide social scripts to follow in specific situations

• Discuss situation ahead of time and make a plan
  ▪ Who will you talk to?
  ▪ What will you say?

• Process situations after the fact and brainstorm solutions
  ▪ What didn’t go well?
  ▪ What else could you have done?
  ▪ What will you do differently next time?

• Formal social skills groups or “lunch bunch”
Emotional and Behavioral EF Interventions

• What EF deficits are contributing to emotional/behavioral difficulties?

• Teach child to ask for and take a time out when needed to leave situation and/or talk with an adult

• Teach an emotional vocabulary
  ● Work to label complex emotions- “Upset” could mean angry, sad, etc.

• Key words and scripts for getting “stuck”- “If I am flexible, more good things happen to me”
  ● Plan A/ Plan B
  ● Big deal/ Little deal
  ● Choice/ No choice
  ● Compromise

• Strategies to increase awareness of problems with emotional regulation
  ● Use “speedometer” to gauge emotion, then tie it to a concrete behavior
John has a book I want

I AM STUCK
Hit John, take book

Time out, miss TV time

Was this what I wanted?

I COMPROMISE
Ask John for a turn

Look at book, give it back, get TV time

Was this what I wanted?

From Kenworthy et al., Solving Executive Functioning Challenges
<table>
<thead>
<tr>
<th>Speed</th>
<th>Level of Anger</th>
<th>Behavior</th>
</tr>
</thead>
<tbody>
<tr>
<td>100</td>
<td>Out of control</td>
<td>Immediately leaving situation</td>
</tr>
<tr>
<td>60</td>
<td>Feeling like I might lose control</td>
<td>Seeking adult help</td>
</tr>
<tr>
<td>30</td>
<td>Getting mad</td>
<td>Counting to delay response</td>
</tr>
<tr>
<td>10</td>
<td>Normal, not mad</td>
<td></td>
</tr>
</tbody>
</table>
From Kenworthy et al., Solving Executive Functioning Challenges
Maintaining Motivation

• Keep it positive (4 to 1)
  ▪ Praise should be specific and true (What creative thought!)
  ▪ “Catch” kids doing something positive

From Kenworthy et al., Solving Executive Functioning Challenges
Maintaining Motivation

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  ▪ “Catch” kids doing something positive

• Use a reward system
  ▪ Internal motivation less helpful for kids
  ▪ “When something is difficult or unexpected, getting bonus points helps to reset my brain”

• Expect more, get more

• Pick your battles

• “Don’t just do something, sit there”

• Keep your sense of humor
**Solving Executive Function Challenges**

Simple Ways to Get Kids with ADHD Unstuck & On Target

Laurie Kniskern, PhD, RERC Research Associate
Karen C. Neale, MEd, RERC Research Associate
Laura M. Eurell, PhD, RERC Coordinator
Leslie Barlow, PhD, RERC Director

**Late, Lost & Unprepared**

Executive Dysfunction & the Disorganized Child/Adolescent

Interventions, strategies and skill building for the child or adolescent who...
- Constantly loses belongings, assignments and directions
- Struggles with organization and time management
- Exhibits disorganized behavior
- Shows poor impulse control
- Does not appear to live up to his/her potential

**SMART but SCATTERED**

Boost Any Child’s Ability to:
- Get Organized
- Resist Impulses
- Stay Focused
- Follow Through
- Solve Problems Independently
- Plan Ahead

Peg Dawson, EdD, and Richard Guare, PhD

**The “Executive Skills” Program for Helping Teens Reach Their Potential**

Over 75,000 in Print!

Richard Guare, PhD, Peg Dawson, EdD, and Colin Guare
Assessment to Assist with Intervention

Psychological Eval
IQ
Academics
Social-Emotional

Neuropsychological Eval
IQ
Academics
Social-Emotional
Language skills
Nonverbal skills
Memory
Attention
Executive functions
Sensory and motor
Using Neuropsych Eval for School

- Helping school to understand educational impact of the medical disorder
  - How this child may be different from other special education students

- Provide recommendations for interventions and accommodations through IEP or 504 Plan
  - IEP
    - Provides accommodations and interventions
    - Child needs to have a specific disability AND it must be "educationally handicapping"
    - Other Health Impaired classification
  - 504 Plan
    - Classroom modifications or accommodations
    - Implementation/guidelines are left up to schools
Take Home Messages

- Children with XY variations at high risk for executive functioning weaknesses, which can impact academic, social, and behavioral functioning.

- Be aware of times of increased risk (certain situations; school transitions).

- Be aware of how others interpret child’s behavior (lazy, not trying) and reframe as EF deficits when appropriate.

- Use “task analysis” to determine where child is having difficulties.

- Use consistent interventions and be patient!
Thank you!

jennifer.janusz@childrenscolorado.org