EXECUTIVE FUNCTIONING IN STUDENTS WITH DOWN SYNDROME

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February 23, 2012
Background: Down Syndrome (DS)

Initially reported by John Langdon Down and 2 others in 1866

Most common chromosomal cause of intellectual disability
Incidence 1: 700
Increased life expectancy, improved quality of life
Behavioral Phenotypes

Behavioral Outcomes
(Cognitive, Linguistic, Social, Emotional, Psychopathology, Motor)

SYNDROME
## Behavioral Phenotypes

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Etiology</th>
<th>Phenotype</th>
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<tbody>
<tr>
<td>Down syndrome</td>
<td>Trisomy 21</td>
<td>Language deficits, social strengths, visual &gt; verbal processing, motor deficits, poor problem solving, task persistence</td>
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<tr>
<td>Williams syndrome</td>
<td>7q deletion</td>
<td>Hypersociability, fears and anxieties, verbal &gt; visuo-spatial, expressive language strengths</td>
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<tr>
<td>Prader-Willi syndrome</td>
<td>15q deletion (paternal)</td>
<td>Mild ID, hyperphagia, hoarding, compulsive behavior, tantrums, skin picking, simultaneous &gt; sequential processing</td>
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<tr>
<td>Smith-Magenis syndrome</td>
<td>17p deletion</td>
<td>Moderate-severe ID, self-injury, hyperactivity, stereotypies, sleep disturbances</td>
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<tr>
<td>5p-syndrome</td>
<td>deletion 5p</td>
<td>Severe ID, atypical catlike cry, hyperactivity, stereotypies, self-injury</td>
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</tbody>
</table>
DS Behavioral Phenotype

**Relative Strengths**
- Visual-spatial processing
- Core social-relatedness
- Receptive language

**Relative Challenges**
- Verbal processing
- Expressive language
- Aspects of motor functioning
- Goal-directed behavior (?)
Cognition in DS

• Cognition = thinking skills, information processing
• What do we currently know about cognition in DS?
  IQ decline during early childhood
  Visual > verbal processing
  Social cognitive abilities
  Challenges with “goal-directed behavior”
Goal-Directed Behavior in DS

Task persistence
(Kasari & Freeman, 2001; Landry & Chapieski, 1990; Pitcairn & Wishart, 1994; Ruskin, Kasari, Mundy & Sigman, 1994; Vlachou & Ferrell, 2000)

More distractible
(Gunn & Cuskelly, 1991)

Stubborn/strong
willed streak
(Carr, 1995; Gibson, 1978)
• Many different approaches to understanding this pattern of difficulty with goal-directedness
• May be helpful to view these patterns within an Executive Function (EF) framework
Executive Functioning (EF)

An umbrella term used to describe the cognitive processes integral to adaptive, goal-directed actions.
Executive Functioning (EF): Components

- Working memory
- Planning
- Shifting
- Inhibition
Significance

**EF skills associated with:**

- Academic readiness and achievement  
  (Blair & Razza, 2007; McClelland et al., 2007)
- Social skills  
  (Diamond, Barnett, Thomas, & Munro, 2007)
- Adaptive behavior  
  (McClelland, Morrison, & Holmes, 2000, Zingerevich & LaVesser, 2009)
- Health related outcomes in TD children  
  (Riggs et al., 2011)
What do we know about EF in DS?

Lack of comprehensive studies, but

• existing literature/preliminary evidence
• marked dissociations even within specific subdomains, such as working memory
EF in DS

• Demonstrated deficits in working memory
  (Jarrold et al., 1999; Lanfranchi et al., 2009)

  intervention work (Conners et al., 2001; 2008)

• Some evidence for deficits in planning
  (e.g. Fidler et al. 2005; Kasari & Freeman, 2001; Rogers et al. 2008)

• Mixed evidence regarding inhibitory control
  (Kopp et al., 1983, Rowe et al. 2006)

• Mixed evidence regarding shifting
  (Edgin 2003; Rowe et al., 2006)
Planning Strategies in DS

DS < DD
Brown-Forsythe’s $F (2,48) = 8.60, p < .001$
Cohen’s $d = 1.18$

Fidler et al. 2005a
Motor Planning

Greater difficulty with motor planning in DS than MA-matched groups:

- coin in the bank, $t(25) = 2.53, p < .01$
- necklace in the cup, $t(25) = 3.45, p < .002$
- pull toy, $t(25) = 2.42, p < .05$
- climb out of the box, $t(25) = 2.15, p < .05$

Fidler et al. 2005b
Research Questions

1. EF in everyday life: do children with DS show difficulties beyond their overall developmental status?

2. Are there some areas of everyday EF skills that are stronger or weaker than others?

Lee, Fidler, Blakely-Smith, Daunhauer, Hepburn & Robinson (2011)
Participants

Completed by 26 parents of children with DS
Children with DS, ages 3-10 yrs drawn from a larger study (DiGuiseppi et al., 2010)
## Participant Characteristics

<table>
<thead>
<tr>
<th></th>
<th>Mean</th>
<th>SD</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronological Age</td>
<td>75.15</td>
<td>23.05</td>
<td>48 - 129</td>
</tr>
<tr>
<td>Mental Age</td>
<td>36.57</td>
<td>8.98</td>
<td>24 - 57</td>
</tr>
<tr>
<td>Developmental Quotient</td>
<td>50.23</td>
<td>9.95</td>
<td>33 - 68</td>
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<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>%</th>
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<tbody>
<tr>
<td>Total sample</td>
<td>26</td>
<td>--</td>
</tr>
<tr>
<td>Female</td>
<td>11</td>
<td>42.3</td>
</tr>
<tr>
<td>White, Non-Hispanic</td>
<td>20</td>
<td>80.0</td>
</tr>
<tr>
<td>Mother Ed, College +</td>
<td>14</td>
<td>58.3</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Father Ed, College +</td>
<td>15</td>
<td>62.5</td>
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1. \( n=25 \)
2. \( n=24 \)
3. Mother’s Education – Number/Percent who completed college
4. Father’s Education – Number/Percent who completed college
Measure

Behavior Rating Inventory of Executive Function – Preschool (BRIEF-P, Gioia, Espy, & Isquith, 2003) 63 Items

Reliability: adequate internal consistency, and test-retest reliability

Validity: Construct and known-group validity reported
BRIEF-P Variables

Index Scales
- Global Executive Composite (GEC)
- Inhibitory Self Control Index (ISCI) = I + EC
- Flexibility Index (FI) = S + EC
- Emergent Metacognition Index (EMI) = WM + PO

BRIEF-P Clinical Domains
- I  Inhibit
- S  Shift
- EC Emotional Control
- WM Working Memory
- P  Plan/Organize
Results: DS T-score means

Lee, Fidler, Blakely-Smith, Daunhauer, Hepburn & Robinson (2011)
Percentage above clinical cut off

Lee, Fidler, Blakely-Smith, Daunhauer, Hepburn, & Robinson, 2011
### BRIEF-P: Working Memory Domain

<table>
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<tr>
<th>Description</th>
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<tr>
<td>When given two things to do, remembers only the first or last</td>
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<tr>
<td>Has trouble completing tasks</td>
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<tr>
<td>Has a short attention span</td>
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<tr>
<td>Has difficulty with activities involving more than one step</td>
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<tr>
<td>Unable to fully describe event, person, or story</td>
</tr>
<tr>
<td>Unaware of his/her [good or poor] performance</td>
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<tr>
<td>Needs adult assistance to stay on task</td>
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<tr>
<td>Forgets what he/she is doing mid-task</td>
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<td>Effort expended on activities falls short of ability</td>
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**BRIEF-P: Plan/Organize Domain**

<table>
<thead>
<tr>
<th>Behavior</th>
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</thead>
<tbody>
<tr>
<td>Puts things away in disorganized, random way</td>
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<tr>
<td>Difficulty beginning and completing tasks</td>
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<tr>
<td>When getting something forgets what he/she is supposed to get</td>
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<td>Can’t find clothes, toys, etc. even with specific instructions</td>
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<tr>
<td>Difficulty following established routines for sleeping, eating, or play</td>
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<td>Difficulty generating solutions or completing an activity when stuck</td>
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<td>Gets distracted by the small details and misses the main idea</td>
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</table>
Summary

3 to 10 yr old children with DS may have significant difficulties in everyday activities requiring specific cool EF functions (working memory and planning)

Lee, Fidler, Blakely-Smith, Daunhauer, Hepburn & Robinson (2011)
NEXT STEPS...
EF Skills & Academic Achievement in DS

In 3 Study Groups: DS, ID, & TD:

1. Characterize the profile of relative strengths and weaknesses in EF in kindergarteners and second-graders

2. Examine how EF skills at Time 1 relate to academic skills and a set of related skills foundational to academic at Time

3. Compare magnitudes of change from kindergarten (Time 1) to second-grade (Time 2) in the development of EF skills across groups

Funded by the FUNDED BY: U.S. DEPARTMENT OF EDUCATION, INSTITUTE OF EDUCATIONAL SCIENCE, SPECIAL EDUCATION RESEARCH GRANTS (R324A110136)
WHAT DO WE DO WITH THIS NEW KNOWLEDGE?
Supporting EF skills in children with DS

- Practicing EF skills in the context of playful activities and games can be beneficial
- New activities and games
- Modifying familiar activities or routines to incorporate practice of EF skills
- Can be imbedded in other therapies (OT, PT, Speech)
Targeting inhibition

• **Games**
  Red light/green light
  Duck, duck, goose

• **Routines**
  Include an element of delay (waiting until everyone is seated to begin eating)
Targeting Working Memory

• **Games**
  Memory/Concentration games
  Simon Says

• **Use organizers**
  Activity Boards
  Picture Schedules

• **Prompts**
  Who goes next in the game?
  What just happened in the story we read?
  Remembering sequences (first hang up your coat, then come and play)
Targeting Shifting

• **Games/activities that have rule switches**
  For example, scan a phonics sheet for a specific letter (“b”) and then search for another letter (h)
  Read joke and riddle books together (especially those with word play)
Acknowledgements

Nancy Raitano Lee, PhD
Susan Hepburn, PhD
Audrey Blakely-Smith, PhD
Cordelia Robinson, RN, PhD
David Most, PhD
Laura Hahn, MS
Julie Hill
Our participants and their families
THANK YOU!
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