EXECUTIVE FUNCTIONING IN DOWN SYNDROME: Implications for education and intervention

Deborah J. Fidler, PhD
Lisa A. Daunhauer, ScD
Human Development & Family Studies Department
Colorado State University
October 19th, 2013
Background: Down Syndrome

Most common chromosomal cause of intellectual disability

Incidence 1: 700

Increased life expectancy, improved quality of life

Initially reported by John Langdon Down and 2 others in 1866
Behavioral Phenotypes

Behavioral Outcomes
(Cognitive, Linguistic, Social, Emotional, Psychopathology, Motor)
Down Syndrome (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?
  - 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 & Cuskelley, 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 (cognitive flexibility)
- Inhibition
- **Working memory** = simultaneously storing and manipulating information for complex tasks such as learning and reasoning (Baddeley, 1992).
- “After you’ve hung up your coats, pick a math station and do two activities” requires working memory.
- **Inhibition**/response inhibition = exerting self-control over behavior to override automatic, or prepotent, responses
- In school: persisting with an assignment despite being irritated by a peer
- (Set-) **Shifting** = cognitive flexibility, the ability to change from one set of rules to another.
- Wisconsin Card Sort Task
- In school: reading directions for a new assignment where one must filter redundancies, focus on salient information and make inferences
Planning = generating appropriate steps needed to reach a goal.

In school: when students successfully prioritize the steps needed to complete an assignment such as a science project.
Blair and Razza (2007): EF assessed in preschool and kindergarten independently predicted math performance, letter knowledge, and phonemic awareness at the end of kindergarten.

McClelland and colleagues (2007): preschoolers (n = 310) with better performance inhibition and working memory at the beginning of the school year had higher scores in literacy, vocabulary, and math at the end of the school year.
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 outcome
  (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 challenges in working memory (Jarrold et al., 1999; Lanfranchi et al., 2009) intervention work (Conners et al., 2001; 2008)
- Some evidence for challenges in planning (e.g. Fidler et al. 2005; Kasari & Freeman, 2001; Rogers et al. 2008)
- Mixed evidence regarding inhibitory control (Kopp et al., 1983, Lee et al. 2011; Pennington et al., 2003)
- Mixed evidence regarding shifting (Edgin 2003; Rowe et al., 2006; Zelazo et al., 1996)
Planning

Object Retrieval Task

Fidler et al. 2005a
Young children with DS show greater difficulty than expected with early problem solving tasks that involve object planning.

\[ DS < DD \]

Brown-Forsythe’s F (2,48) = 8.60, p < .001

Cohen’s d = 1.18

Fidler et al. 2005a
Motor Planning

Young children with DS show greater difficulty with motor planning than expected for overall developmental level on praxis tasks:

- 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?
3. Do parents and teachers report similar profiles of everyday EF function?
4. What is the relative risk for executive difficulties?
5. Does EF relate to functional performance of activities in everyday life?

Daunhauer, Fidler, Hahn, Lee, & Hepburn (2013)
Lee, Fidler, Blakely-Smith, Daunhauer, Hepburn & Robinson (2011)
Procedures

- Teacher-reported data
- Parent-reported data
- Direct child assessment

A component of two larger projects examining EF in school-aged children with DS
Measures

<table>
<thead>
<tr>
<th>Instrument</th>
<th>Citation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Behavior Rating Inventory of Executive Function – Preschool</td>
<td>BRIEF-P</td>
</tr>
<tr>
<td></td>
<td>Gioia et al., 2003</td>
</tr>
<tr>
<td>Leiter-Revised</td>
<td>Leiter-R</td>
</tr>
<tr>
<td></td>
<td>Roid &amp; Miller, 1995; 1997</td>
</tr>
<tr>
<td>School Function Assessment</td>
<td>SFA</td>
</tr>
<tr>
<td></td>
<td>Coster et al., 1998</td>
</tr>
<tr>
<td>Pediatric Evaluation of Disability Inventory</td>
<td>PEDI</td>
</tr>
<tr>
<td></td>
<td>Haley et al., 1992</td>
</tr>
</tbody>
</table>
BRIEF-P Variables

**Index Scales**

- Global Executive Composite \( \text{GEC} \)
- Inhibitory Self Control Index \( \text{ISCI} = I + EC \)
- Flexibility Index \( F_I = S + EC \)
- Emergent Metacognition Index \( \text{EMI} = WM + PO \)

**BRIEF-P Clinical Domains**

- I \( \text{Inhibit} \)
- S \( \text{Shift} \)
- EC \( \text{Emotional Control} \)
- WM \( \text{Working Memory} \)
- P \( \text{Plan/Organize} \)
## EF: DS, TD Teacher-Reported Findings

<table>
<thead>
<tr>
<th>Domain</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>DS group &gt; TD</strong></td>
<td></td>
</tr>
<tr>
<td>Global Exec Composite</td>
<td>$F(1, 33) = 13.43, p = .001$</td>
</tr>
<tr>
<td>Inhibitory Self-Control</td>
<td>ns</td>
</tr>
<tr>
<td>Flexibility Index</td>
<td>ns</td>
</tr>
<tr>
<td><strong>DS group &gt; TD</strong></td>
<td></td>
</tr>
<tr>
<td>Emergent</td>
<td>$F(1, 33) = 21.91, p &lt; .001$</td>
</tr>
<tr>
<td>Metacognition</td>
<td></td>
</tr>
<tr>
<td>Inhibit</td>
<td>ns</td>
</tr>
<tr>
<td>Emotional Control</td>
<td>ns</td>
</tr>
<tr>
<td>Shift</td>
<td>ns</td>
</tr>
<tr>
<td><strong>DS group &gt; TD</strong></td>
<td></td>
</tr>
<tr>
<td>Working Memory</td>
<td>$F(1, 36) = 26.68, p \leq .001$</td>
</tr>
<tr>
<td>Plan/Organize</td>
<td>$F(1, 36) = 8.09, p = .007$</td>
</tr>
</tbody>
</table>

Note: Using Bonferroni adjustments
In other words...

- Teachers reported greater difficulty (than expected for overall developmental status) in the areas of **working memory** and **planning** in DS.
Teacher BRIEF-P Reports:
% Clinically Elevated [T score > 65]

BRIEF-P Domains

BRIEF-P: ↑ scores, ↑ EF symptoms
In other words...

- The majority of children with DS showed clinically high levels of difficulties in the areas of working memory and planning, according to teachers.
Parent BRIEF-P Reports:
% Clinically Elevated [T score > 65]

<table>
<thead>
<tr>
<th>BRIEF-P Domains</th>
<th>Total Indexes</th>
<th>Scales</th>
</tr>
</thead>
<tbody>
<tr>
<td>GEC</td>
<td>*</td>
<td></td>
</tr>
<tr>
<td>ISCI</td>
<td>*</td>
<td></td>
</tr>
<tr>
<td>FI</td>
<td></td>
<td></td>
</tr>
<tr>
<td>EMI</td>
<td>*</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>*</td>
<td></td>
</tr>
<tr>
<td>EC</td>
<td></td>
<td></td>
</tr>
<tr>
<td>S</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WM</td>
<td></td>
<td>*</td>
</tr>
<tr>
<td>P/O</td>
<td></td>
<td>*</td>
</tr>
</tbody>
</table>

BRIEF-P: ↑ scores, ↑ EF symptoms
In other words...

- According to parents, majority of children with DS showed clinically high levels of difficulties in the areas of **working memory** and **planning**

- A small subgroup also show clinically high levels of difficulties with **inhibition**
# BRIEF-P: Working Memory Domain

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

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

<table>
<thead>
<tr>
<th>DOMAIN</th>
<th>$r$</th>
<th>$p$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Global Executive Composite</td>
<td>.73</td>
<td>.001</td>
</tr>
<tr>
<td>Emergent Metacognition Index</td>
<td>.50</td>
<td>.030</td>
</tr>
<tr>
<td>Flexibility Index</td>
<td>.64</td>
<td>.003</td>
</tr>
<tr>
<td>Inhibitory Self Control Index</td>
<td>.69</td>
<td>.001</td>
</tr>
</tbody>
</table>
In other words...

- Parents and teachers agreed most of the time on these dimension ratings, but not always
Relative Risk (RR) of Developing EF Challenges?

<table>
<thead>
<tr>
<th>BRIEF-P Scale</th>
<th>RR</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Working Memory</td>
<td>6.70</td>
<td>3.61-12.44</td>
</tr>
<tr>
<td>Plan/Organize</td>
<td>5.26</td>
<td>2.68-10.35</td>
</tr>
<tr>
<td>Inhibit</td>
<td>3.43</td>
<td>1.49-7.54</td>
</tr>
</tbody>
</table>
What is school function?

- School function involves “a student’s ability to perform important functional activities that support or enable participation in the academic and related social aspects of an educational program”
- (Coster et al., 1998, p 2).
School function

Examples:

- using school-related materials appropriately (such as writing tools and books),
- the ability to move around the school environment independently,
- the ability to manage self-care and personal needs,
- requesting assistance when needed (Coster et al., 1998)
School function

- School function and the academic aspects of schooling are different.
- Academic skills involve class instruction and homework involving acquisition of knowledge on specific content areas such as reading and math.
- With foundational school function skills, students are able to engage in academic-related activities without the need for assistance or accommodations.
What factors are important for adequate school function in DS?
# Predictors of School Function

<table>
<thead>
<tr>
<th>Variable</th>
<th>B</th>
<th>SE B</th>
<th>B</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>MA (Leiter-R)</td>
<td>-.30</td>
<td>.33</td>
<td>-.18</td>
<td>.385</td>
</tr>
<tr>
<td>Language Development (OWLs)</td>
<td>-.04</td>
<td>.23</td>
<td>-.03</td>
<td>.867</td>
</tr>
<tr>
<td>EF- Teacher Report (BRIEF-P Global Composite)</td>
<td>-.86</td>
<td>.19</td>
<td>-.74</td>
<td>\leq .001</td>
</tr>
<tr>
<td>Constant</td>
<td>134.34</td>
<td>20.32</td>
<td>\leq .001</td>
<td></td>
</tr>
</tbody>
</table>

Note: Adjusted $R^2 = .55; F = (3, 17), p \leq .001$
## Functional Performance (PEDI) and EF

<table>
<thead>
<tr>
<th>DOMAIN</th>
<th>( r )</th>
<th>( p )</th>
</tr>
</thead>
<tbody>
<tr>
<td>Working Memory-Self Care PEDI</td>
<td>.60</td>
<td>( \leq .01 )</td>
</tr>
<tr>
<td>Plan/Organize-Self Care PEDI</td>
<td>.56</td>
<td>( \leq .01 )</td>
</tr>
<tr>
<td>Working Memory-Caregiver Assistance PEDI</td>
<td>.63</td>
<td>.003</td>
</tr>
<tr>
<td>Plan/Organize-Caregiver Assistance PEDI</td>
<td>.64</td>
<td>.001</td>
</tr>
</tbody>
</table>
In other words...

Executive function skills (especially planning and working memory) are critical for both school function and functioning in daily life.
Next steps...
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)
- Anticipatory guidance
Targeting Working Memory and Planning

- **Games**
  Memory/Concentration games, computer games
  Simon Says

- **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 inhibition

**Activities**
- Red light/green light
- Duck, duck, goose
- Yoga, martial arts, other mindful practices

**Routines**
- Include an element of delay (waiting until everyone is seated to begin eating)

**Strategies**
- Ask child to verbalize problem-solving strategies
Targeting Shifting/Cognitive Flexibility

- **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)

- **Exercise!**
  Aerobics, sports
Curricula

- Tools of the Mind
- PATHS
- Montessori
- Chicago School Readiness Project
Pairing and Sharing

- Brainstorming “take aways”
  - Applications to your life?
  - Activities and strategies you are already using?
Example Resources

  http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3159917/  
- SuperFlex Curriculum  
  http://www.socialthinking.com/?view=featured
Funding Acknowledgements

**EF Skills & Participation in Everyday Life in DS**

H133G100197
U.S. Department of Education, National Institute of Disability and rehabilitation research
THE NATIONAL INSTITUTE OF DISABILITY AND REHABILITATION RESEARCH (NIDRR)

**EF Skills & Academic Achievement in DS**

R324A110136
U.S. Department of Education, institute of educational science, Special Education Research Grants
Acknowledgements

Collaborators

- Wendy J. Coster, PhD
- Nancy Raitano Lee, PhD
- Susan Hepburn, PhD
- Audrey Blakely-Smith, PhD
- Cordelia Robinson, RN, PhD
- David Most, PhD
- Jeannie Visootsak, MD

DD Research Lab Team

Graduate Research Assistants
- Brianne Gerlach-McDonald
- Elizabeth Will, M. Ed.
- Amber Lopez

Research Assistants
- Chelsea Fimia-Moe
- Aubree Goudy
- Olivia Onofrio
- Kristen Stejskal
- Lauren Whynott

Alumni
- Laura Hahn, PhD
Acknowledgements: Our Participants

THANK YOU!
Contact:
Lisa.Daunhauer [at] colostate.edu
Deborah.Fidler [at] colostate.edu

DD Research Lab http://ddlab.colostate.edu/