SUPPORTING THE DEVELOPMENT OF EXECUTIVE FUNCTION IN YOUNG CHILDREN WITH DOWN SYNDROME

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DOWN SYNDROME BEHAVIORAL PHENOTYPE

Relative strengths:
• Receptive Language
• Visual processing
• Social Relatedness*

Relative challenges:
• Expressive Language
• Verbal Processing
• Motor Development
• Self-regulation
  – Goal-Directed Behavior
  – Executive Function
OVERVIEW OF PRESENTATION

Developmental Research in Down syndrome

Links to Outcomes in home, school, community

Translation to Practice
Why study developmental processes in neurogenetic syndromes?
Developmental dynamics approaches focus not only on endstates, but on how an individual progressively develops and reaches various endstates throughout the lifespan

(Fidler, Lunkenheimer, Hahn, 2011)
Developmental dynamics approach to behavioral phenotypes can:

- Help researchers formulate interventions that are time-sensitive
- Intervene to support adaptive developmental cascades
DEVELOPMENT OF COGNITIVE PHENOTYPE IN DS

Infant profile: Foundations of Goal-Directed Behavior

Toddler profile: Early Goal-Directed Behavior

Middle Childhood and beyond: Executive Function
• Are early cognitive developmental features detectable in toddlers with DS?

• Is it possible to characterize differences in rates of growth in areas of strength and vulnerability?

*Fidler et al., 2005a; b, Fidler et al., 2006; Fidler et al., 2008a*
EMERGING PHENOTYPE IN DS

Cross-sectional findings in toddlers with DS (Fidler, Hepburn & Rogers)

- Participants
  - 18 toddlers with Down syndrome
  - 19 toddlers with mixed/non-specific developmental delays
  - 24 MA-matched typically developing infants and toddlers

- Disability groups equated on CA ($M = 33$ mos)

- All groups equated on MA ($M = 21$ mos) gender
EMERGING PHENOTYPE IN DS

• All children had normal vision or vision corrected to within normal range
• No hearing impairments beyond those commonly associated with Down syndrome
• No impaired hand use, all were mobile
• No between disability group differences on premature births, degree of early intervention services received
• Parents of typical children were significantly younger, but no Educational or SES differences
Emerging Phenotypic Profile

Mullen Scales of Early Learning

Vineland Adaptive Behavior Scales

Fidler et al, 2006
Object retrieval

- 15 trials
- Prize placed under a plexiglass box with one opening, child instructed to retrieve the prize through the opening
- Coded for quality of “retrieval strategy”
EARLY DEVELOPMENT IN DS

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

Fidler et al. 2005a
EARLY PLANNING SKILLS *(FIDLER ET AL. 2005A)*

- Children with Down syndrome were more likely to produce less optimal strategies:
  - attempt to reach through the top of the plastic box (where there was no opening) $F(2, 48) = 2.88, p < .05$
  - look through the side opening of the box to locate the toy, straighten up, and then reach appropriately $F(2, 48) = 5.59, p < .001$
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, $t(25) = 2.15, p < .05$
EARLY PLANNING SKILLS (FIDLER ET AL., 2005A)

• Coins in the bank:
  – 91.7% unable to plan through holding coins in their hand, and then transferring a coin from the palm of their hand to their fingers, and then to the bank

• Pull toy:
  – 72.7% could not coordinate walking multiple steps and watching the toy at same time
Preliminary evidence for emerging difficulties with goal-directed behavior and object-related planning
DEVELOPMENTAL DYNAMICS

• Is it possible to detect the emerging profile in a more dynamic way?

• Can we detect the emergence of this profile from infancy into toddlerhood?
EMERGING DS PHENOTYPE (FIDLER ET AL., 2008)

- NICHD Early Childcare Study/Collaboration with Univ. of Washington (Cathryn Booth-LaForce, Jean Kelley)
- Down syndrome $n = 23$; idiopathic DD $n = 25$
- Bayley Scales of Infant Development-II at 12 and 30 months
- Both groups matched for nonverbal mental age at 12 mos (Bayley raw score = 70)
Bayley Mental Scale Age Equivalent: 12 & 30 months

- Bayley Mental Scale Age Equivalent: 12 & 30 months

- Fidler et al., 2008

- Bayley 12 mo. MA
- Bayley 30 mo. MA

- DD
- DS
BAYLEY II ITEMS

• Mental Scale 6-12 mos
  • Prefers novel visual display
  • Plays with rattle
  • Reaches for ring
  • Looks for fallen spoon
  • Rings bell purposely
  • Lifts cup by handle

• 15-20 mos
  • Retrieves toy (clear box)
  • Places peg in pegboard
  • Six beads in a box
  • Finds one object
  • Removes object from bottle
  • Places three cubes in cup
BAYLEY ORIENTATION AND ENGAGEMENT SCALES

Fidler et al., 2008b
ORIENTATION/ENGAGEMENT SCALE

- Orientation to examiner
- Social engagement
- Cooperation
- Attempts to interact socially
- Trusting the examiner/lack of fearfulness
EMERGING DS PHENOTYPE (FIDLER ET AL., 2008)

• Early social relatedness skills emerge with greater competence from 12 to 30 months in young children with DS

• Early cognitive foundations emerge with less competence from 12 to 30 months in young children with DS
How does this profile continue to emerge and develop over time?
EXECUTIVE FUNCTION IN DOWN SYNDROME

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EXECUTIVE FUNCTION

Cognitive Processes integral to adaptive and goal directed behavior

Working Memory, Inhibitory Control, Cognitive Flexibility (Shifting), and Planning
EXECUTIVE FUNCTION: COMPONENT PROCESSES

- **Working Memory**: Holding and manipulating information while completing a task
  - Ex: Multi-step Instructions

- **Inhibitory Control**: Control of pre-potent responses
  - Ex: Raising Hand

- **Cognitive Flexibility**: Ability to modify thinking and/or strategies across contexts
  - Ex: Changing behavior based on setting

- **Planning**: Means-end behavior reliant on some or all of the other constructs
  - Ex: Completing work for recess

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Will, Fidler, Daunhauer, & Gerlach-McDonald (2017)
EXECUTIVE FUNCTION

Health Outcomes

Adaptive Behavior

Social Skills

Academic Readiness

Academic Achievement

Executive Function

(Blair & Razza, 2007; Bornstein et al., 2013; Diamond et al., 2007; McClelland et al., 2000; 2007; Riggs, 2012)
EF MEASUREMENT

Proxy Report

Laboratory Assessment
EF RESEARCH I: LABORATORY BASED MEASURES
**WORKING MEMORY/INHIBITION**

**Pony & Gator**
- Form of Simon Says
- Child must remember rule to respond correctly
- Language check
- 4 practice trials
- 10 experimental trials
- Coding: Total correct responses
- Kappa $M = .89; .62 - 1.0$
INHIBITORY CONTROL

Snack Delay

- Snack placed under cup
- Retrieve snack when bell rung
- 4 trials:
  - 5-, 10-, 15-, 20-s
- Coding: Total disinhibited behaviors
- Kappa $M = .78; .61-.94$
DCCS

- Cards sorted on what shape dimension
- 1 practice
- 6 trials first rule
- Rule is changed
- 6 trials post-switch
- 12 trials border version
- Coding: Total correct responses
- Kappa $M=.85; .64-1.0$
PLANNING

Generativity
-Children are told Here are some toys for you
-Play with unusual group of objects
-2 minutes
-Coding: Total new objects explored; total functional acts
-Kappa $M = .77$; $.63 - 1.0$
METHODS

**DS**

N = 42
NVMA = 49.98 mos
CA = 91.07 mos

**TD**

N = 38
NVMA = 50 mos
CA = 40.52 mos

Daunhauer, Gerlach-McDonald, Will & Fidler (2017)
## RESULTS

<table>
<thead>
<tr>
<th></th>
<th>DS n= 42</th>
<th>TD n= 38</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M (SD)</td>
<td>M (SD)</td>
<td></td>
</tr>
<tr>
<td>Raw Score</td>
<td>.591(2.17)</td>
<td>7.16(2.39)</td>
<td></td>
</tr>
<tr>
<td>Z-Score</td>
<td>-.29(.83)</td>
<td>.23(.92)</td>
<td></td>
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<tr>
<td>Working memory</td>
<td></td>
<td></td>
<td>.05</td>
</tr>
<tr>
<td>Inhibition</td>
<td>.76(1.26)</td>
<td>.23(.77)</td>
<td>.131</td>
</tr>
<tr>
<td>Shifting</td>
<td>.495(.44)</td>
<td>.68(.42)</td>
<td>.163</td>
</tr>
<tr>
<td>Planning</td>
<td>5.10(2.41)</td>
<td>7.0(3.0)</td>
<td>.01</td>
</tr>
</tbody>
</table>
EF STUDIES II: ECOLOGICAL MEASURES OF EF

Teacher-reported data
Parent-reported data

DS = 25; TD = 23 (NVMA)
## EF STUDIES I: ECOLOGICAL MEASURES OF EF

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Down syndrome $n = 25$</th>
<th>Typically developing $n = 23$</th>
<th>$t$/$X^2$</th>
<th>$p$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronological age (months)</td>
<td>96.56 ± 17.31 61–133</td>
<td>39.78 ± 5.00 30–46</td>
<td>15.71</td>
<td>.001</td>
</tr>
<tr>
<td>Mental age (months)$^a$</td>
<td>50.12 ± 8.32 40–67</td>
<td>49.96 ± 5.32 35–57</td>
<td>.22</td>
<td>.83</td>
</tr>
<tr>
<td>Child’s gender female</td>
<td>7 f 25 n 28.00 %</td>
<td>9 f 23 n 39.10 %</td>
<td>0.19</td>
<td>.89</td>
</tr>
<tr>
<td>Child White, Non-Hispanic</td>
<td>22 24 91.70 %</td>
<td>19 23 82.60 %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mother ed, college$^b$</td>
<td>14 25 68.00 %</td>
<td>21 23 91.30 %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mother White, Non-Hispanic</td>
<td>23 24 96.00 %</td>
<td>19 23 82.6 %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Father ed, college$^b$</td>
<td>17 24 71.00 %</td>
<td>21 23 91.30 %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Father White, Non-Hispanic</td>
<td>23 23 100 %</td>
<td>21 23 91.30 %</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

$^a$Leiter-R Brief IQ raw score used for statistical comparison. $^b$Mother and Father’s Education – Number/Percent who completed college.
PARENT BRIEF-P REPORTS:
% CLINICALLY ELEVATED [T SCORE > 65]

BRIEF-P Domains

BRIEF-P: ↑ scores, ↑ EF symptoms
IN OTHER WORDS...

According to parents, clinically high levels of difficulties in the areas of working memory and planning.
TEACHER BRIEF-P REPORTS:
% CLINICALLY ELEVATED [T SCORE > 65]
EARLIER WORKSHOP

• How do EF skills relate to school performance in the primary grades in DS?
• How do EF skills relate to adaptation in home, work, and community settings in DS?
INFANT FOUNDATIONS OF GOAL-DIRECTED BEHAVIOR: EARLY PLANNING SKILLS
INFANT FOUNDATIONS OF GOAL-DIRECTED BEHAVIOR

What infant foundations are necessary for competence in goal-directed behavior?
Earliest goal-directed behaviors involve reaching and grasping objects for exploration
FOUNDATIONS OF GOAL-DIRECTED BEHAVIOR

Motor Development

Visual Attention to Objects

Motor Cognition

Early Goal-Directed Behavior with Objects
DISCUSSION

How can syndrome-related findings help shape early intervention planning and programming?

What are some ways that you may already be supporting EF?
1. Syndrome-informed intervention *planning*
   - Anticipatory guidance
2. Syndrome-specific intervention *techniques*
   - target emerging areas of strength and challenge with new interventions

*Fidler, Philofsky & Hepburn (2007)*
SYNDROME-INFORMED INTERVENTION PLANNING

- Behavioral phenotype research may make it possible to “know where to look” for potential vulnerabilities.

- Parents/practitioners use information regarding phenotypic predispositions to monitor potential areas of vulnerability.
Anticipatory Guidance:

- anticipate potential developmental vulnerabilities and resiliencies
- Select interventions that will target areas of subtle vulnerability before they become pronounced areas of weakness/delay

*(Fidler, Philofsky & Hepburn, 2007)*
SYNDROME-INFORMED INTERVENTION PLANNING (FIDLER, PHILOFSKY & HEPBURN, 2007)

If interventions are natural and without harm, then potential “over-employment” is of low risk, while the gains may be of high benefit.
Second approach:

Craft new, targeted interventions specific for observed phenotypic profile
Fey et al: certain interventions have been shown to be specifically ineffective for subgroups of children with DD
Responsive Education/Prelinguistic Milieu Teaching- RE/PMT

Children with DDs responded with greater gains in instrumental (goal-directed) requesting than children in the DS group.

DS group not receiving RE/PMT actually demonstrated faster growth in requesting skills than DS intervention group.

(Yoder & Warren, 2002)
EXECUTIVE FUNCTION

Targeting the emerging phenotype in young children with DS
PHENOTYPE-INFORMED INTERVENTION

• Anticipatory Guidance:
  – Monitor the development of early nonverbal communication skills in young children with DS, especially as children reach overall MA of 9 months
  – Monitor the development of early goal-directed behavior, problem solving, motor planning, non-verbal requesting
General Speech-Language Intervention can:

- Encourage requesting through naturalistic opportunities
- “communicative temptations” (Prizant & Wetherby, 1999)
• General Occupational Therapy Intervention can provide opportunities for:
  – early motor exploration (infancy),
  – functional play with objects (toddlerhood),
  – tool use (toddlerhood), and
  – chaining behaviors toward a goal
• Can we craft interventions that will target these skills specifically for young children with DS?

• DD Lab@CSU:
  
  B-BOPS Intervention (Building Blocks of Problem Solving)
  
  • Early strategizing
  • Nonverbal requesting
  • Goal-directed chains of behavior
B-BOPS INTERVENTION

- Requesting
  - Promote use of a range of strategies (gives, reaches, points, eye gaze)
- Goal-directed chains of behavior
  - Games that require 2, 3, 4 steps for a child to reach a desired goal
- Inhibitory control
  - Embed waiting routines into activities
  - Successful strategy is not the prepotent strategy
Promoting Goal Directed Behavior in Infants with DS

- Facilitation of reaching and grasping
- Enhancing object interest
- Promotion of early acts on objects
GENERAL IMPLICATIONS

• Early emergence of behavioral phenotypes: should we wait for areas of strength and weakness to become pronounced?

• Bridging behavioral phenotype science and intervention practice will strengthen early intervention for children with neurogenetic syndromes
  – Identify subtle manifestations of later, more pronounced outcomes
  – Monitor and target those areas in an active and informed way
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QUESTIONS?
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