

THE
2ND DECADE



MYOTONIC
DYSTROPHY
FOUNDATION



2017 MDF Annual Conference

Sept. 8–9, 2017 San Francisco



MYOTONIC
DYSTROPHY
FOUNDATION

Care and a Cure

THE PSYCHOLOGICAL COMPLEXITY OF MYOTONIC DYSTROPHY

Missy Dixon, PhD, MS



HEALTH
UNIVERSITY OF UTAH

Introduction

- **DM1 affects more than physical function**
- **Psychological component**
 - Cognitive functions
 - Psychosocial functioning
- **Why is this important?**
 - Impacts affected individual, family, and caregiver
 - Physical changes may contribute to psychological changes
 - Foster healthy social and emotional growth

Defining the Psychobable

- **Cognitive Functioning-** means and mechanisms of acquiring knowledge (i.e., reasoning, memory, perception, awareness, attention, judgment, and language)
- **Executive Function-** cognitive processes necessary for the cognitive control of behavior
- **Psychosocial Functioning-** psychological function of individual in the context of his/her social environment
- **Behavioral Functioning-** behavior in the context of social environment

Physical Concerns

- **Progressive muscle weakness**
- **Slow motor development**
- **Tire easily**
- **Sit to stand difficulties**
- **Decreased muscle power**
- **Myotonia** and locking up
- **Falls**
- **GI**
- **Eating and Swallowing**

Implications

- **Loss of muscle and strength**
 - Fatigue
 - Tired
 - Falls
 - Anxiety
 - Depression
- **Physical symptoms that may impact psychological functioning**
 - Fatigue, sleep problems, incontinence, GI complaints, pain, headache, swallow
 - May contribute to anxiety, depression, attention, memory, daily functioning, executive function problems

Addressing Physical Concerns

- **Communication**
 - ▣ Express concerns and feelings
 - ▣ Active listening
- **Adapt environment to disease progression**
 - Adapted environment contributes to feelings of success and not feelings of loss
 - Increase social interactions
 - Decrease apathy, depression, anxiety
- **Adaptive devices, power chairs, AFOs**
 - Sense of freedom and stability
 - Increase confidence and social interactions
 - Less fearful of falling

Cognitive Function

- **Thinking**
- **Memory**
- **Language**
- **Attention**
- **Executive Function**
 - ▣ Planning
 - ▣ Organization
 - ▣ Inhibition
 - ▣ Shift
 - ▣ Emotional control
 - ▣ Initiation
 - ▣ Working memory
 - ▣ Monitoring behavior

Research

□ **Health Endpoints and Longitudinal Progression in Congenital Myotonic Dystrophy (HELP-CDM)**

- Nicholas Johnson, MD, University of Utah
- Craig Campbell, MD, Western University, Canada
- 3 year study following 50 children with CDM
- Cognitive assessments: IQ, executive function, adaptive behavior, autism, QOL, sleep

□ **Health Endpoints and Longitudinal Progression in Myotonic Dystrophy (HELP-DM)**

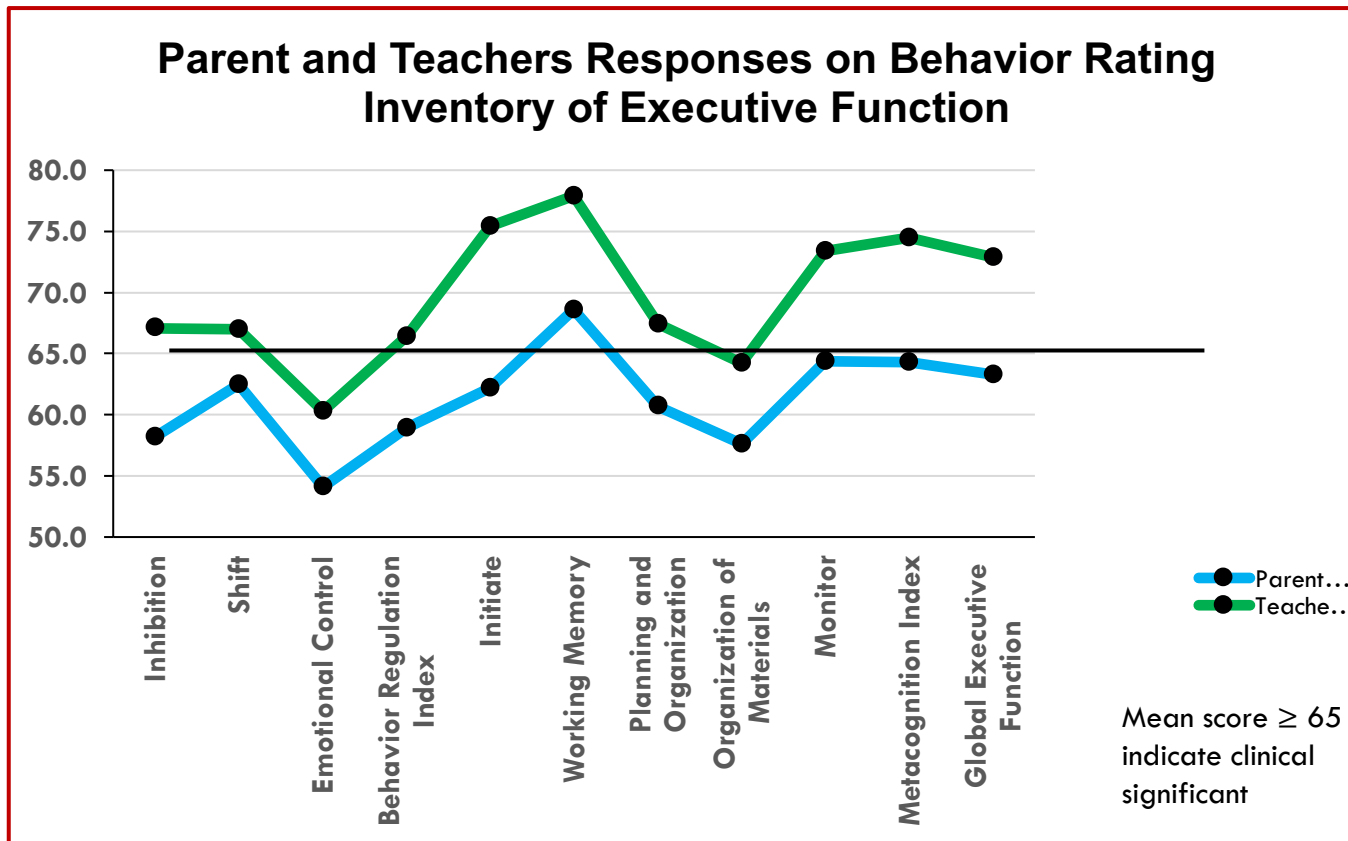
- Nicholas Johnson, MD, University of Utah
- 1 year study with f/u at 3 months of 22 adults with DM1
- Cognitive assessments: IQ, executive function, adaptive behavior, memory, visuospatial abilities, processing speed, QOL, sleep, anxiety, depression, apathy

What did we learn from HELP-CDM?

Cognitive Functions: CDM vs Control

Construct	Measure	CDM \bar{x} (sd)	Control \bar{x} (sd)	t-score (p-value)
Intelligence (IQ)	<i>WPPSI-III/WAIS-IV</i>	66.1 (18.1)	100 (15)	-10.60 (0.000)
Adaptive Behavior	<i>Vineland-II</i>	70.0 (16.2)	125.6 (15.5)	-10.15 (0.000)
Executive Function	<i>BRIEF parent</i>	63.3 (11.1)	≤ 65	6.20 (0.000)
	<i>BRIEF teacher</i>	72.9 (13.8)	≤ 65	6.80 (0.000)
Autism Traits	<i>ASSQ</i>	15.0 (9.3)	≤ 19	6.10 (0.000)
Social Communication	<i>SCQ</i>	12.4 (7.1)	≤ 15	5.33 (0.000)
Repetitive Behavior	<i>RBS-R</i>	16.4 (14.7)	≤ 13	NA

What did we learn from HELP-CDM?



What did we learn from HELP-DM?

- ▣ IQ: $x = 88.15$ (9.7), average (norm 85-115)
- ▣ BDI: $x = 11.68$ (7.4), moderate depression
- ▣ BAI: $x = 11$ (8.0), moderate anxiety
- ▣ EF: Shift $x = 67$ (WM & initiate- trending)
- ▣ Slowed processing speed
- ▣ Memory impairment

What does this look like in daily life?

- **EF/CF problems affect language, memory, and concentration**
 - Automation, planning, and learning
 - Task completion
 - Time orientation
 - Initiative
 - Resistance to change
 - Switching tasks
 - Behavior
 - Mood

Important to know

- **Cognitive function issues are not one of physical function or motivation.**
- **Individuals are not unwilling.**
- **Unable to complete tasks and follow directions in the same way as others.**
- **Rethink the way we approach giving tasks/asking for things.**

What can you do to help?

- **Strategize to find ways that work for individuals**
- **Memory: short sentences, clear instructions, divide information**
- **Visual cues to help process information**
- **Check in- Does she understand?**
- **Let him repeat what he's been told to synthesize and build verbal memory**
- **Summarize and repeat information**
- **Introduce new information in steps**

What can we do to help?

- **Extra time for instructions**
- **Break tasks into smaller parts (two 10 minute vs. one 20 minute)**
- **Quiet location with limited interruptions**
- **Make lists and check off completed tasks (individuals can do)**
- **Agenda training (individuals can plan their days, weeks, etc.)**
- **Extra time for transitions**
- **Help get things started**
- **Create a system for organizing and planning**
- **Use lists and calendars**
- **Create system with check lists for self-monitoring**

Important to know

- **Emotional and behavioral problems are normal to healthy socio-emotional development**
- **Emotional reaction to disease progression is normal for affected individuals and family**
 - Stages of grief
 - Denial
 - Anger
 - Bargaining
 - Depression
 - Acceptance

Psychosocial Functioning

- Emotional and behavioral concerns
 - ▣ Important clinically because children/adults recognize consequences of their disease
- Psychosocial adjustment related to:
 - Relationships w/ friends (↓ w/ age and/or disease progression)
 - Dependence, hostility, productivity (↑↓ w/ disease progression)
 - Anxiety, depression, and withdrawal (↑ w/ age or disease progression)

Psychosocial Functioning

- **Learned helplessness:** when an action does not have an impact, inclination to give up hope and make no further effort
 - Can lead to
 - emotional reaction (apathy and depression)
 - behavior problems (passivity, tantrums)
- **How to help**
 - Activities that individual can complete (create a positive experience)
 - Increased sense of control leads to increased motivation to continue

Psychosocial Functioning

□ Emotional concerns

- Increase when symptoms increase; associated anxiety
- Increase when there is a loss of independence and/or control-
 - response: hostility, depression, relief
- Increase when peers are moving on and individuals feel stuck because of their DM

□ How to help

- Important to respond as you would to any child/adult
- Normalize emotional response given circumstances
- Mourn the loss to help move toward acceptance
 - Stages: denial, anger, bargaining, depression, acceptance
- Address emotions, learning opportunity, listen, name emotions, solve problems and set limits
- Work together to be successful!

Psychosocial Functioning

- **Tantrums/Wallowing**
 - ▣ Normal process of growing up
 - ▣ Want 100% of your attention
 - ▣ Best to address tantrum with boundaries
- Normal course of tantrum: 1) reason no matter how small or meaningless, 2) escalation, 3) explosion, 4) cooling down
- **How to help**
 - ▣ Set limits, respond clearly, allow tantrum to run the course w/o intervention or reaction, time out to cool down, discuss emotions
 - ▣ Accept person's emotions, but set limits on negative behavior

Caregivers burn-out is a real thing

- Take time for yourself-this is not being selfish
 - Relaxation, sleep, exercise
 - Date nights
 - Time outs

**You are a better caregiver when you
take time for yourself!**

Resources

- Myotonic Dystrophy Foundation
 - www.myotonic.org
- MDA
 - www.mda.org

Acknowledgements

University of Utah

Nicholas Johnson, MD

Russ Butterfield, MD, PhD

Kiera N. Berggren, MA, MS

Brith Otterud

Rebecca Crockett

Brandon Zielinski, MD, PhD

Man Hung, PhD

Jerry Bounsaga

Research participants and families

MDF/Wyck Foundation Fellowship

MDA

Valerion Therapeutics

University of Rochester

Heather Adams, PhD

Western University

Craig Campbell, MD

Karen Bax, PhD



Questions?
