MEANINGFUL AND SAFE ENGAGEMENT IN THE COMMUNITY FOR PERSONS AND FAMILIES WITH HUNTINGTON’S DISEASE
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- University of Virginia (UVA) Health System
- UVA HD Clinic: HDSA Center of Excellence
Presenter Disclosures

K. M. “Shelley” Knewstep-Watkins, OTR/L

The following personal financial relationships with commercial interests relevant to this presentation existed during the past 12 months:

No relationships to disclose or list
Objectives

– Describe the physical, psychological and cognitive symptoms

– Describe how the symptoms impact a person’s community-based activities

– Identify OT intervention for working, driving, social participation, and leisure
INTRODUCTION/REVIEW: HUNTINGTON’S DISEASE
HD is Rare

- 30,000 people with HD
- 200,000 people who are at risk for HD

(HDSA, 2016a)
What is HD?

- Neurodegenerative
  - Progressive
- Inherited
  - Autosomal dominant
    - Each Child = 50/50 chance
  - A family disease
What is HD?

Mutated Huntingtin Gene
(≥36 CAG repeats)

Mutated Huntingtin Protein

Mutated Protein is Toxic to Brain Cells

(Nance, 2011); (Rosenblatt, 2011); (HDSA, 2016a)
When is it HD?

• Gene status determined before birth but diagnosed clinically by symptoms

• # of CAG repeats explains ~60-70% of the variation in age that symptoms begin
  – ↑ CAG repeats correlates to earlier symptoms

• Typical onset of HD symptoms: age ~35-55
  – >90% do not have predictive testing

(Nance, 2011); (Rosenblatt, 2011)
Clinical Presentation

- Cognitive Impairments
- Psychological Impairments
- Physical Impairments

Recommended: Search “Daniel My Brother (Huntington’s Disease)” on YouTube for a brief clip of an inspirational person and family sharing about HD symptoms
Clinical Presentation

- Cognitive Impairments
- Psychological Impairments
- Physical Impairments

Physical Impairments
Clinical Presentation

- Cognitive Impairments
- Psychological Impairments
- Physical Impairments

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- Psychological Impairments
- Physical Impairments
Clinically Defining HD

2 Scales

1. Unified Huntington’s Disease Rating Scale (UHDRS)

2. The 3 Clinical Stages: Early, Middle, Late
The Unified Huntington’s Disease Rating Scale (UHDRS)

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<thead>
<tr>
<th>Domain</th>
<th>Ability</th>
<th>Score</th>
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<tr>
<td>Occupation</td>
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<tr>
<td></td>
<td>Marginal work only</td>
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</tr>
<tr>
<td></td>
<td>Reduced capacity for usual job</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Normal</td>
<td>3</td>
</tr>
<tr>
<td>Finances</td>
<td>Unable</td>
<td>0</td>
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<tr>
<td></td>
<td>Major assistance</td>
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</tr>
<tr>
<td></td>
<td>Slight assistance</td>
<td>2</td>
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<tr>
<td></td>
<td>Normal</td>
<td>3</td>
</tr>
<tr>
<td>Domestic Chores</td>
<td>Unable</td>
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<tr>
<td></td>
<td>Impaired</td>
<td>1</td>
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<tr>
<td></td>
<td>Normal</td>
<td>2</td>
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<td>Activities of Daily Living</td>
<td>Total care</td>
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<tr>
<td></td>
<td>Gross tasks only</td>
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</tr>
<tr>
<td></td>
<td>Minimal impairment</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Normal</td>
<td>3</td>
</tr>
<tr>
<td>Care level</td>
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</tr>
<tr>
<td></td>
<td>Home for chronic care</td>
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<td>Home</td>
<td>2</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>Range 0 - 13</strong></td>
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Shoulson and Fahn Staging Scale

<table>
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<th>TFC Total Score</th>
<th>Stage</th>
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<tbody>
<tr>
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<tr>
<td>7 - 10</td>
<td>II</td>
</tr>
<tr>
<td>3 - 6</td>
<td>III</td>
</tr>
<tr>
<td>1 - 2</td>
<td>IV</td>
</tr>
<tr>
<td>0</td>
<td>V</td>
</tr>
</tbody>
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Least Impaired

(Rosenblatt, 2011)
3 Clinical Stages

• Early Stage HD
  – *May* continue to work, drive, handle money
  – *May* continue to live independently
  – *Possible* symptoms:
    • minor involuntary movements, slight loss of coordination
    • difficulty with complex thinking
    • depression, irritability, disinhibition

• Middle Stage HD
• Late Stage HD

Summarized from: (Rosenblatt, 2011, p.7)
HD Prodrome

• Or “premanifest”

• Time period for a person who is gene-positive for HD before person is clinically-diagnosable

• Clinical and biological traits of HD can occur ~15 years before presenting the signs and symptoms required for the clinical diagnosis of HD

(Paulsen et al., 2010)
The Care Team

Local
- Family, Caregivers
- Primary Care Doctor
- Neurologist (?)
- Counselors, trainers, teachers, etc. (?)
- Therapists – OT, PT, SLP (?)

HD Clinic
- Family, Caregivers
- Neurologist
- Genetic Counselor
- Genetic Counselor
- Psychologist and/or Psychiatrist
- Social Worker
- Therapists – **OT**, PT, SLP
OT & HDSA Centers of Excellence

- 39 HDSA Centers of Excellence (COE) in 30 states
  - Find the closest one to you on the HDSA.org website!
- Clinic visits: ~1-2x/year

A few of UVA HD Clinic’s team members at HDSA Convention 2016
OT Over the Course of the Disease

HD Clinic OT (1-2x/yr.)

Early Stage HD

Outpatient OT (incl. CDRS OT)

Middle Stage HD

Home Health OT

Late Stage HD

Acute Care OT

SNF OT

Person with HD

Recommendation:
OT to OT Collaboration!
OT for Persons and Families with HD

A perfect fit!
EVIDENCE-BASED PRACTICE:
RELEVANT RESEARCH
Paulsen et al. (2014) investigated the predictors of clinically diagnosable HD

- Strongest predictors of diagnosis of HD:
  - Change in motor score, imaging, & cognition score

- Measurable changes in brain structure, cognitive ability, and motor ability before the HD diagnosis

- “the diagnosis of [HD] is made fairly late in the disease course”
Paulsen et al. (2010): exposed decline in IADLs during prodrome

- More likely to remain in typical job IF scored better on:
  - cognitive assessment (SDMT)
  - BDI-energy
  - UHDRS total motor score

- 1st areas of functional decline: work & financial capacity

- Functional decline in select IADLs can occur before the clinical symptoms required for diagnosis of HD

(Paulsen et al., 2010, p. 2599)
Research for OTs

Cruickshank et al. (2015) studied the impact of multidisciplinary rehab for 15 persons with manifest HD

- Intervention: 9-month long multidisciplinary program including fortnightly OT
  - OT focused on cognition & executive function

- Participants demonstrated:
  - improved verbal learning and memory
  - increased grey matter volume

- Participants showed improvements while participating in multidisciplinary intervention (including OT)

(Cruickshank et al., 2015, p. 6).
McCabe, Roberts, & Firth (2008) qualitatively studied impact of progressive neurological illness on work and leisure

**WORK:**
- >80% of patients: full-time work → unemployment
- 51% of caregivers moved to part-time work
- ~1/3 of patients indicate work changes negatively impacted social lives: social isolation, limited funds to socialize, reduced self-confidence

**LEISURE:** Patients’ recreational activities deteriorate due to illness

(McCabe, Roberts, & Firth, 2008)
OT PRACTICE:
FOR PERSONS & FAMILIES
Building Blocks for OT Intervention

Client-Centered and Occupation-Oriented
Building Blocks for OT Intervention

Recognize all aspects of the domain of OT

Client-Centered and Occupation-Oriented

Knewstep-Watkins
Building Blocks for OT Intervention

- Recognize progressive nature
- Recognize *all* aspects of the domain of OT
- Client-Centered and Occupation-Oriented
Building Blocks for OT Intervention

- Establish rapport founded in trust and respect
- Recognize progressive nature
- Recognize *all* aspects of the domain of OT
- Client-Centered and Occupation-Oriented
Building Blocks for OT Intervention

- Primary caregivers present
- Establish rapport founded in trust and respect
- Recognize progressive nature
- Recognize *all* aspects of the domain of OT
- Client-Centered and Occupation-Oriented

Knewstep-Watkins
Building Blocks for OT Intervention

Supportive Environment

Primary caregivers present

Establish rapport founded in trust and respect

Recognize progressive nature

Recognize *all* aspects of the domain of OT

Client-Centered and Occupation-Oriented
OT for the Person with HD

OT Model of Practice?
OT: The Person-Environment-Occupation Model

- **Person**
- **Environment**
- **Occupation**

**Impairments**:
- Cognitive Impairments
- Physical Impairments
- Psychological Impairments

**Occupational Performance**
Occupational Therapy for People with Huntington’s Disease: Best Practice Guidelines

Created by:
The OT Standards of Care working group of the European Huntington’s Disease Network

Published: 2014

(Cook, Page, Wagstaff, Simpson, & Rae, 2014)
Core Concepts for Independence

- A Consistent Daily Routine
- Quiet Environment
- Tidy and Well-Organized Environment
- An Accessible Environment
- Family, Friend, and/or Caregiver Support for Modifications
EMPLOYMENT

Loss of Job

Difficulty at Work
Cognitive? Physical? Behavior?

Leaving On “My” Terms

OT can be a valuable part of the team to address function in the workplace!
EMPLOYMENT

Sample Interview:

– Is your job causing you stress lately?
– Any negative feedback from your supervisor?
– Does anyone at work know you have HD? How would they respond?
– What type of relationship do you have with your co-workers/supervisor?

– Financial Planning? Insurances?
  • Health insurance, life insurance, disability
  • Strongly Recommend:
    Social worker at a HDSA COE or through HDSA
EMPLOYMENT

1st: Collaborate with patient, family, and social worker
2nd: Determine Rehabilitative vs. Compensatory

Rehabilitative Approach
• Consider **Vocational Rehabilitation** (state program)

Compensatory Approach
• Most commonly used
• General recommendations
EMPLOYMENT:
General Compensatory Recommendations

• Reduce distractions
• Adjust work duties
• Adjust work hours
• Reduce commute
• Change jobs

• Additional Considerations:
  – Types of Harm
  – Burden of Stress

List of Vocational Rehab by State:
http://askjan.org/cgi-win/TypeQuery.exe?902
Volunteering

The just-right-fit may be volunteering!

Avoid roles that are: detail-oriented, fast-paced, or with variable conditions

Examples:
• Volunteering at a SPCA
• Volunteering for a church or civic organization
• Being involved in your local HDSA Chapter
I have spread my dreams under your feet;
Tread softly because you tread on my dreams.

W.B. Yeats

Safe driving:
• Complex task that requires reliable:
  ✓ cognitive abilities
  ✓ motor abilities
  ✓ visual processing
  ✓ control of one’s behavior
DRIVING

• Ask the person with HD and his/her family:
  - any recent tickets or accidents
  - any dents or scratches in the vehicle
  - any concerns

• Eventually, person with HD will have to retire from driving
  ➢ Early dialog: “How do you want to make the decision to retire from driving?”
  ➢ Best plan for this person with HD and family?
Mostly common approaches to retire from driving:

1. Incident: Accident, Injury, DMV Safety Report
2. Personal/Family Decision with Clinic Rec.
3. Certified Driver Rehabilitation Specialist (CDRS)
DRIVING: CDRS

An evaluation by CDRS removes the pressure and blame from being on family members and caregivers, and it allows a professional to guide the difficult decision.

“We want to enable you to drive.”

Clay Huie, OTR/L, CDRS
DRIVING: CDRS

An in-vehicle driving evaluation by a CDRS - the safest choice!

• Finding a CDRS:
  – “Search for provider” on ADED website

• Other Considerations?
  – Referral?
  – Cost: *Typically* out-of-pocket
  – Variables: Whose vehicle? Where?
DRIVING: Personal/Family Decision with Clinic Recommendations

• If CDRS too expensive, in-clinic communication among providers, patient, and family/caregivers is critical
  – Resources:
    • Free Home & Care Safety Guides by The Hartford
    • “We Need to Talk: Family Conversations with Older Drivers”

• Strongly recommend:
  Monthly ride-alongs by trusted, responsible adult
DRIVING: Compensatory Strategies

- Maintain extra space around the vehicle
- Minimize distractions
- Do not speed
- Avoid higher speed roads
- Avoid congested traffic
- Avoid cruise-control

Help vs. Harm:
- GPS
- Handicap-accessible parking
DRIVING: Personal/Family Decision with Clinic Recommendations

• Person with HD no longer safe?
  – Family members/caregivers and/or healthcare provider(s) must address; Collaborate

• Doctor’s order for the person with HD to stop driving

• Confidential Reporting to DMV (i.e. Virginia’s Medical Review Request)
Do the activities that can be enjoyed now, NOW!
If you can enjoy travelling now, go now!
If you like to do family hiking trips and can enjoy doing that now, GO DO IT NOW!
LEISURE and SOCIAL PARTICIPATION

✓ 1st: Get to know them! What do they enjoy?
✓ 2nd: Collaborative problem solving and sharing among OT, person with HD, and their family/friends.
✓ 3rd: Encourage them to try and then CELEBRATE the effort and success!
SOCIAL PARTICIPATION

Sources of anxiety/stress:
• Changes in routine
• Unfamiliarity
• Social groups

Strategies to minimize anxiety/stress:
• Consistently in routine
• Discussing social plans in advance
• Writing plans on visible calendar
• “Comfortable” people
LEISURE

• Leisure occupations:
  – Socializing
  – Relaxing
  – The pursuit of interests or hobbies

• Role of the OT: “Ensure that the activity is adapted to fit the person and not the other way around”

(Blacker, Broadhurst, & Teixeira, 2008)
Always an athlete, Joe remains active – staving off the physical effects of Huntington’s. “We bike about four days a week and play some golf,” says Joe. Recently, he took up yoga. “It’s OK with me if I am the worst in my yoga class, as long as I am working on my breathing, balance, and flexibility.” And he challenges [his wife] on the Wii Fit ski slopes.

. . . “I respect my limitations, but I’m not letting them keep me from doing what I love.” (Dubuque, 2015)
LEISURE & SOCIAL PARTICIPATION

Reader:
✓ Try short-stories, magazines, and newspapers.

TV/Movie Watcher:
✓ Try shorter and/or recorded TV shows or watching sports.

Social Butterfly:
✓ Try socializing through texting or e-mail.
LEISURE

Outdoorsman
• Avoid high risk situations
  (e.g. hunting, fishing on a boat, etc.)

Gardeners
• Try a container and/or patio garden.
LEISURE: Exercise

Walkers and Runners
- Always carry cell phone
- Recommend community track
LEISURE: Exercise

Resources:

HDSA: HD Workout Tips by Shana Verstegen

The Huntington Society of Canada
Li (2013) article
• Tai Chi beneficial for people with movement disorders

Evan Cameron’s Insights on Tai Chi for Persons with HD
• Encourages body control and body awareness
• Movements help keep strength and flexibility
• Focuses on breathing and syncing [breathing] with movement
• Focus is on the fundamentals of Tai Chi, like weight shifting, balance, and body control

(Li, 2013); (personal communication, April 25, 2016)
LEISURE and SOCIAL PARTICIPATION

• Bikers:
  • Try helpful features: wider tires, step-through frame, 3-wheeled bike, or recumbent bike.
  • Avoid vehicular traffic and riding alone

Daisy – The cutest dog ever!
“It's funny, my memory is terrible; my time awareness is shot; my concentration is short; but as long as I do my art in short spurts, it works. Although I can't do armatures like I used to because it takes logical thinking ahead and I can't do that anymore.”
LEISURE and SOCIAL PARTICIPATION: VACATIONING

Keep it simple and predictable to keep it FUN!
OT for Persons and Families with HD

A perfect fit for early intervention for community-based occupations!
A Sincere Thanks
References


Li, F. (2013). Tai Ji Quan Exercise for People with Parkinson’s Disease and Other Neurodegenerative Movement Disorders. International Journal of Integrative Medicine, 1(4), 1-5.


Interesting Correlation!

Persons with HD “with more passive lifestyles experienced onset of the illness 4.6 years earlier than those people who were more active”

(Cook et al., 2014, p. 10)