



**MEANINGFUL AND SAFE  
ENGAGEMENT IN THE COMMUNITY  
FOR PERSONS AND FAMILIES WITH  
HUNTINGTON'S DISEASE**

# Shelley Knewstep-Watkins OTR/L

- University of Virginia (UVA) Health System
- UVA HD Clinic: HDSA Center of Excellence



## PATIENT SERVICES

- SERVICES
- PATIENTS & VISITORS
- DOCTORS
- LOCATIONS
- APPOINTMENTS & REFERRALS
- CLINICAL TRIALS & RESEARCH
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FIND A LOCATION

### Huntington's Disease Clinic



University Hospital  
Primary Care Center  
Fourth Floor  
1221 Lee St.  
Charlottesville, VA 22908

**Phone:** 434.924.8668  
**Fax:** 434.982.0606

**Location Hours:**  
Monday - Friday | 8:00 a.m. - 5:00 p.m.

#### LOCATION MAP AND DIRECTIONS



# 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?

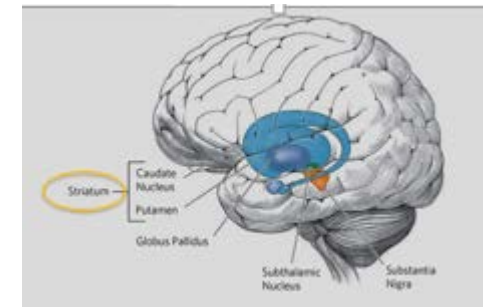
**Mutated Huntingtin  
Gene**  
( $\geq 36$  CAG repeats)



**Mutated  
Huntingtin  
Protein**



**Mutated Protein  
is Toxic to Brain  
Cells**



(Nance, 2011); (Rosenblatt, 2011); (HDSA, 2016a)

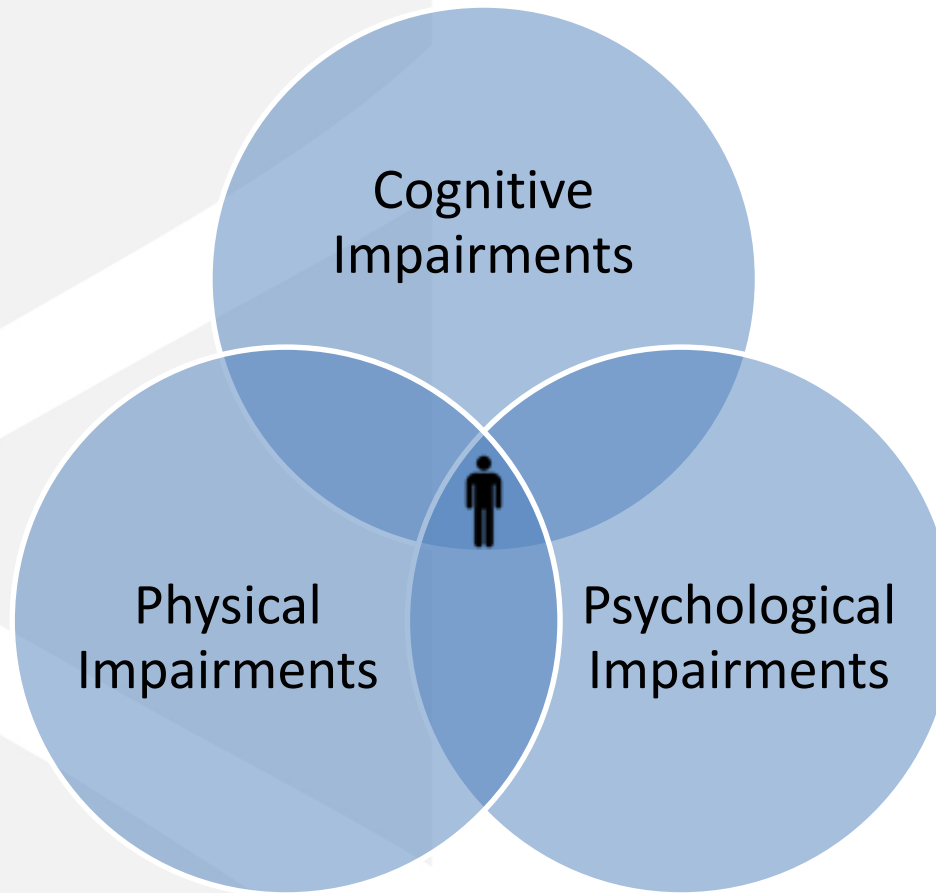


# When is it HD?

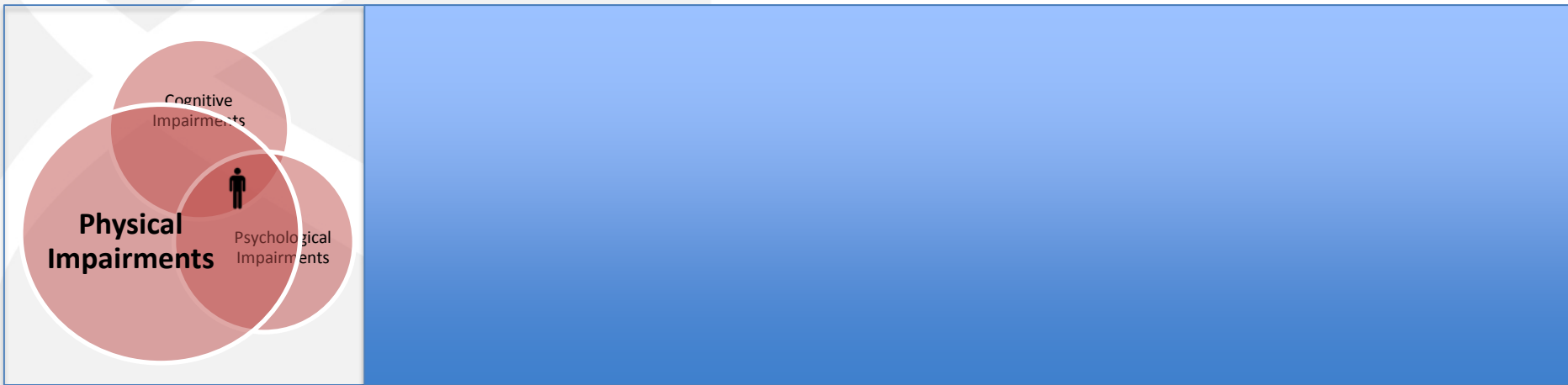
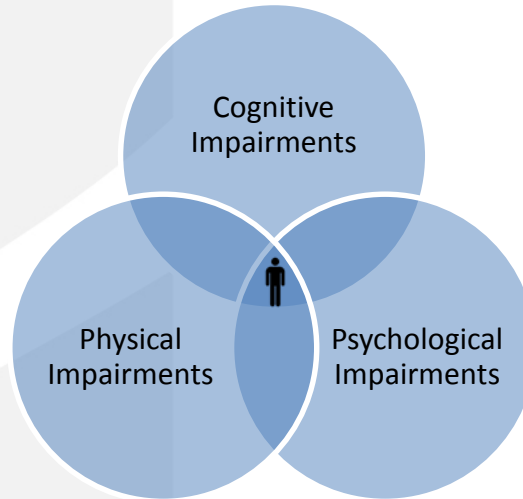
- **Gene status determined before birth *but* diagnosed clinically by symptoms**
- # of CAG repeats explains ~60-70% of the variation in age that symptoms begin
  - ↑ CAG repeats correlates to earlier symptoms
- **Typical onset of HD symptoms: age ~35-55**
  - >90% do not have predictive testing

(Nance, 2011);  
(Rosenblatt, 2011)

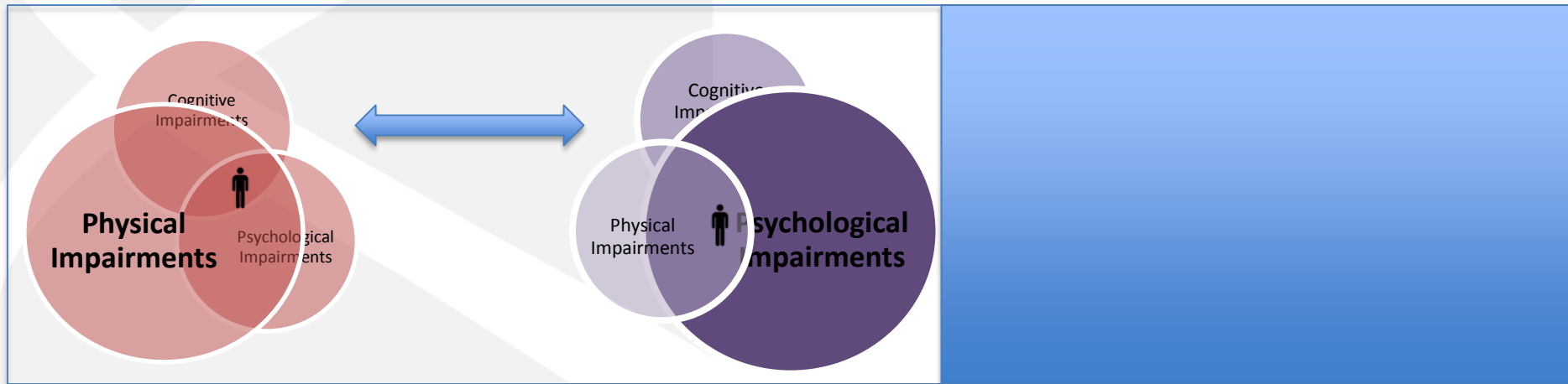
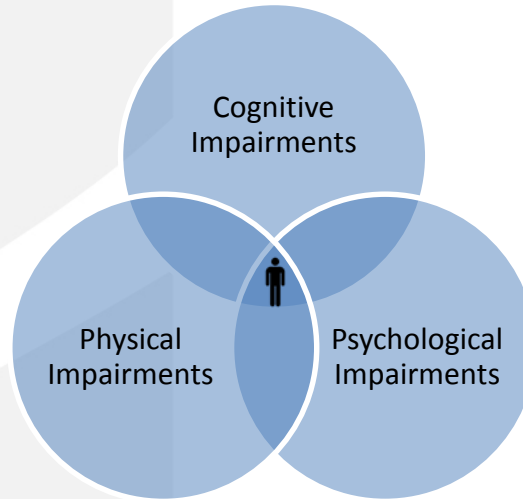
# Clinical Presentation



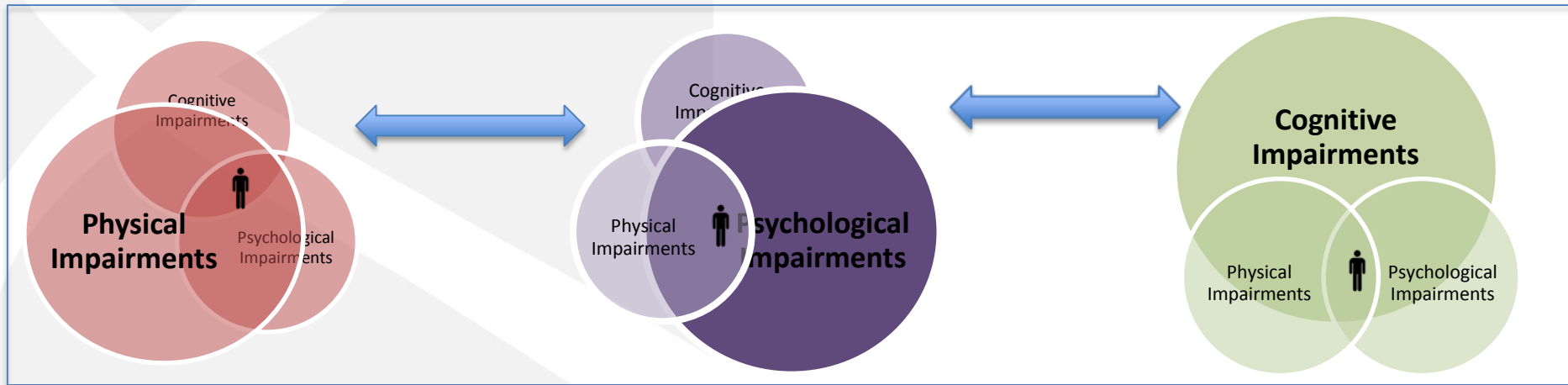
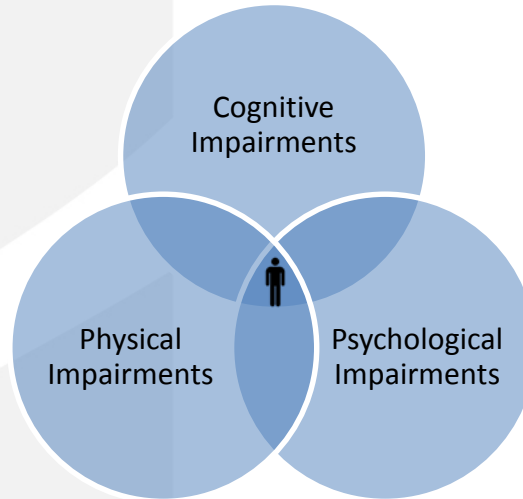
# Clinical Presentation



# Clinical Presentation



# Clinical Presentation



# 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)

Total Functional Capacity Rating Scale <sup>1</sup> (Source: UHDRS <sup>2</sup> )		
Domain	Ability	Score
Occupation	Unable	0
	Marginal work only	1
	Reduced capacity for usual job	2
	Normal	3
Finances	Unable	0
	Major assistance	1
	Slight assistance	2
	Normal	3
Domestic Chores	Unable	0
	Impaired	1
	Normal	2
Activities of Daily Living	Total care	0
	Gross tasks only	1
	Minimal impairment	2
	Normal	3
Care level	Full-time nursing care	0
	Home for chronic care	1
	Home	2
<b>TOTAL</b>	Range 0 - 13	

*Shoulson and Fahn Staging Scale<sup>1</sup>*

TFC Total Score	Stage
11 - 13	I
7 - 10	II
3 - 6	III
1 - 2	IV
0	V

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

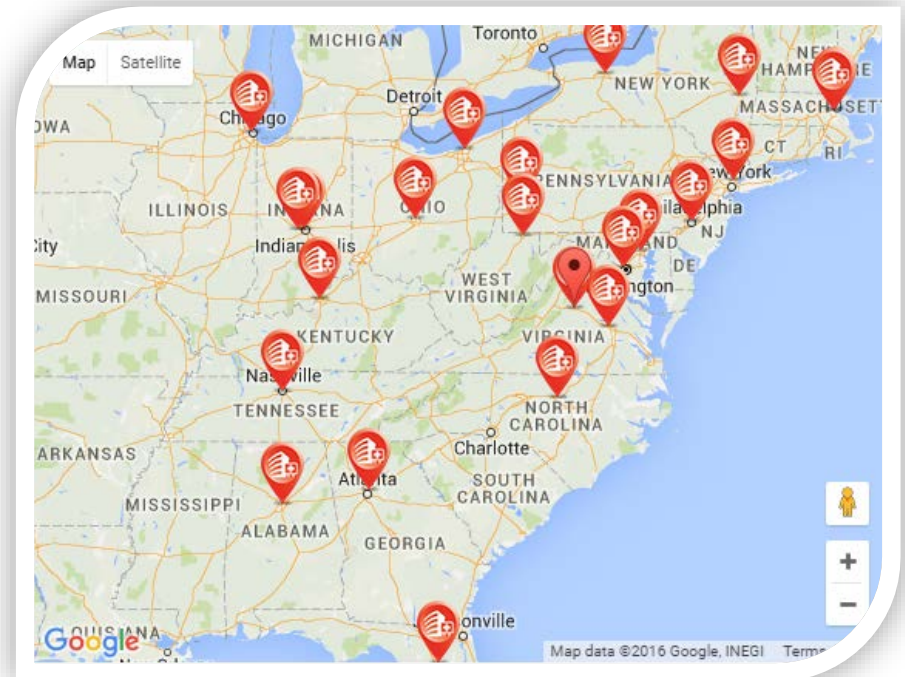
- Family Caregivers
- Neuropsychologist
- Genetic Counselor
- Psychologist and/or Psychiatrist
- Social Worker
- Therapists – OT, PT, SLP

# OT & HDSA Centers of Excellence

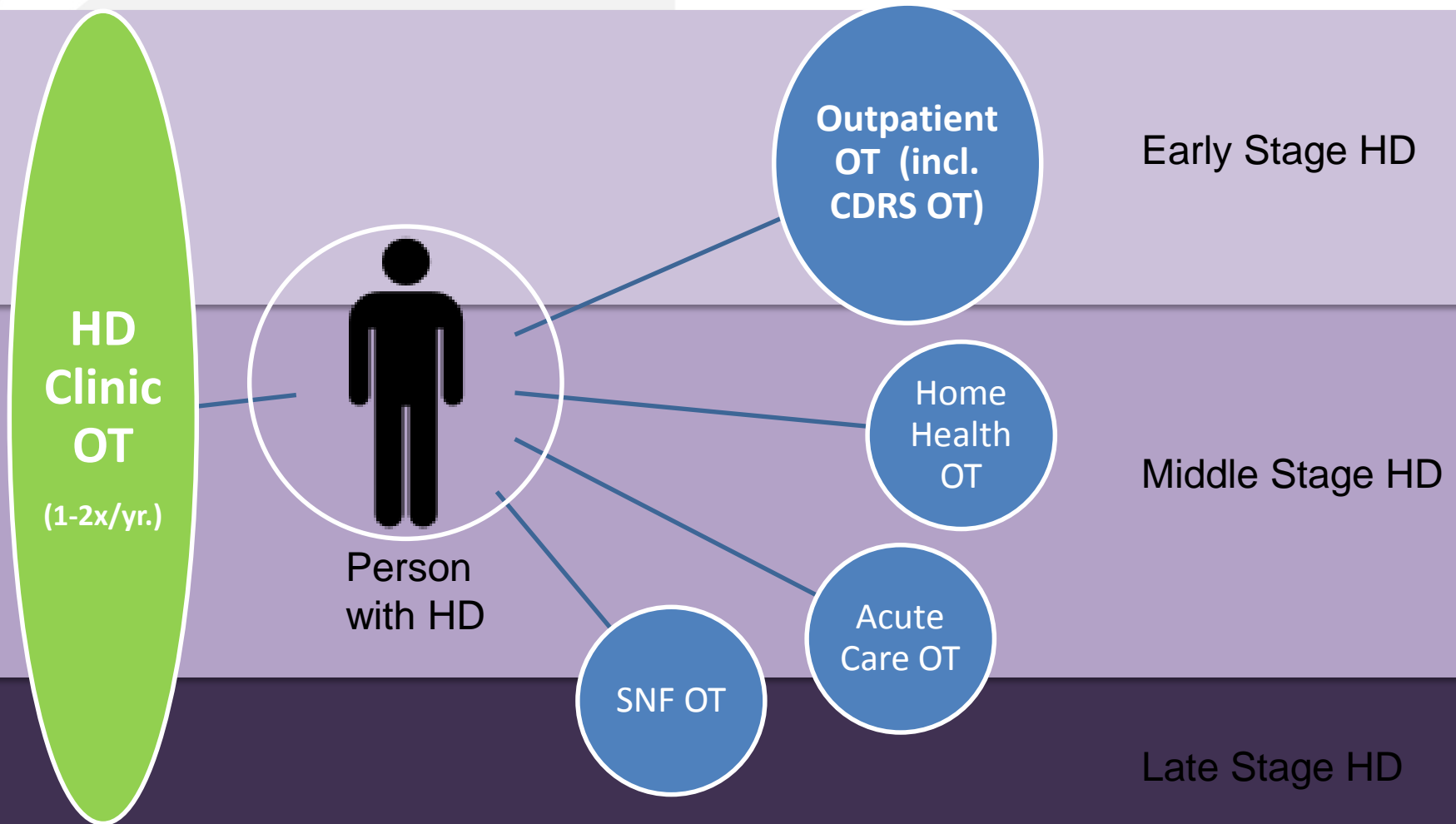
- 39 HDSA Centers of Excellence (COE) in 30 states
  - *Find the closest one to you on the [HDSA.org](http://HDSA.org) website!*
- Clinic visits: ~1-2x/year



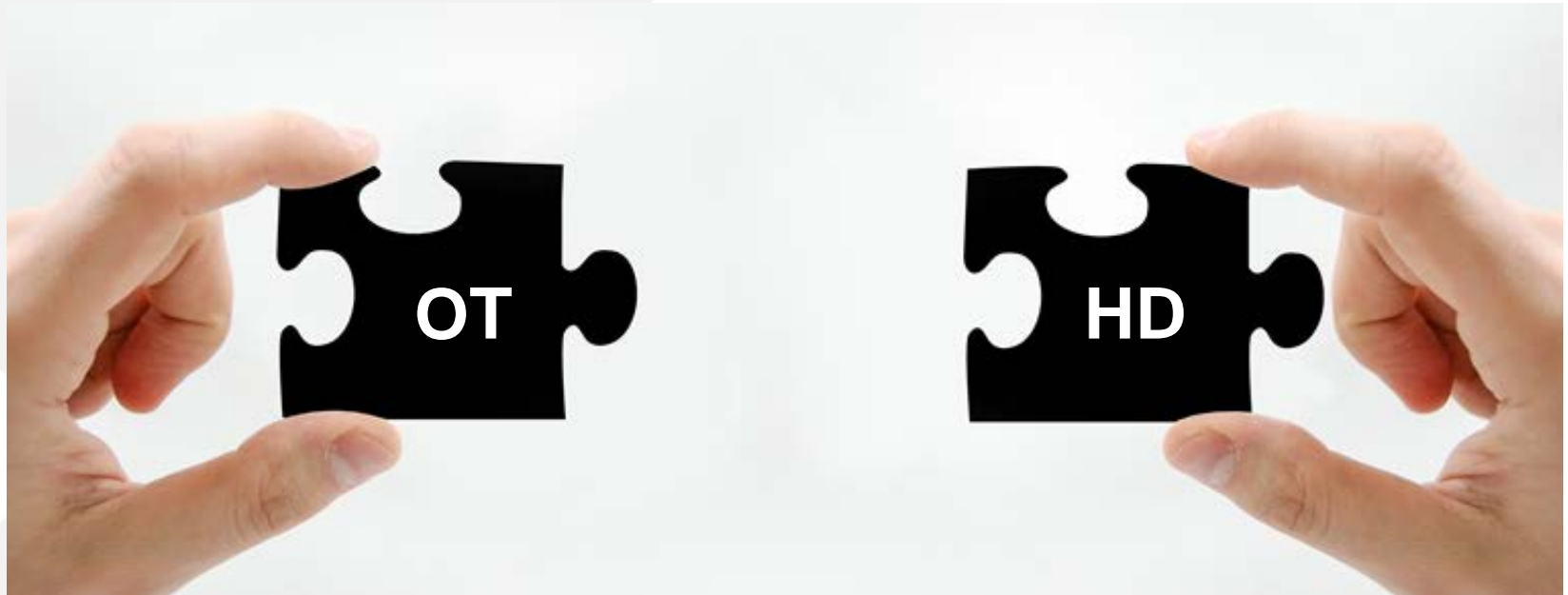
*A few of UVA HD Clinic's team members at HDSA Convention 2016*



# OT Over the Course of the Disease



## OT for Persons and Families with HD



**A perfect fit!**

# **EVIDENCE-BASED PRACTICE: RELEVANT RESEARCH**

## Research for OTs

**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., 2014)

## Research for OTs

**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
- 1<sup>st</sup> 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).

## Research for OTs

**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

*Mostly  
negative  
feelings!*

- ~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**

Knewstep-Watkins

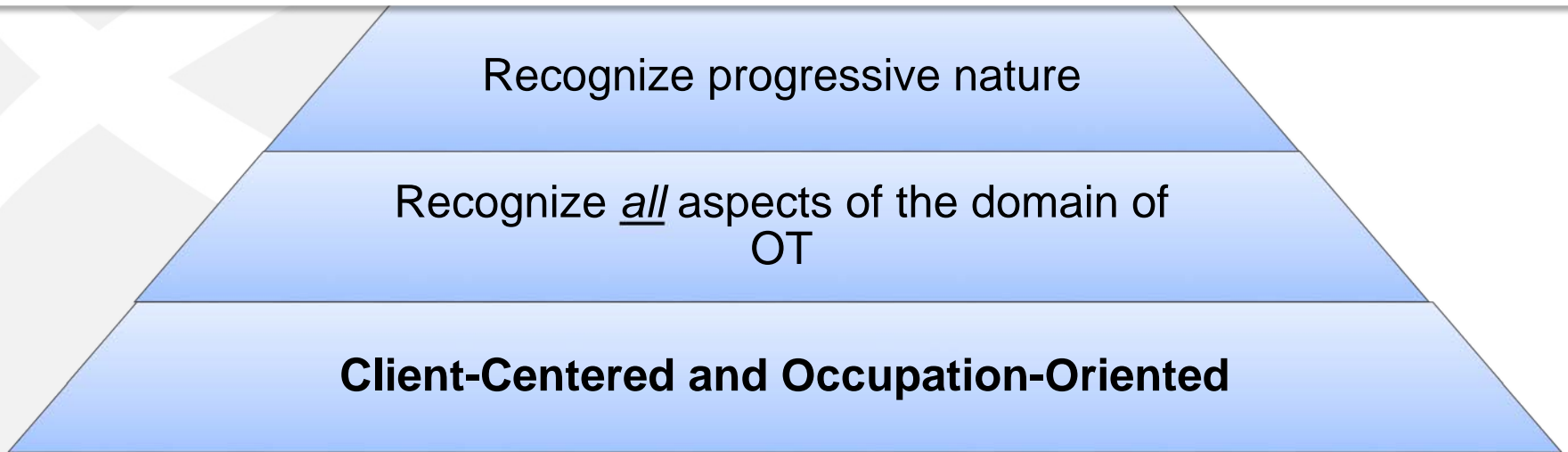
# Building Blocks for OT Intervention

Recognize all aspects of the domain of  
OT

**Client-Centered and Occupation-Oriented**

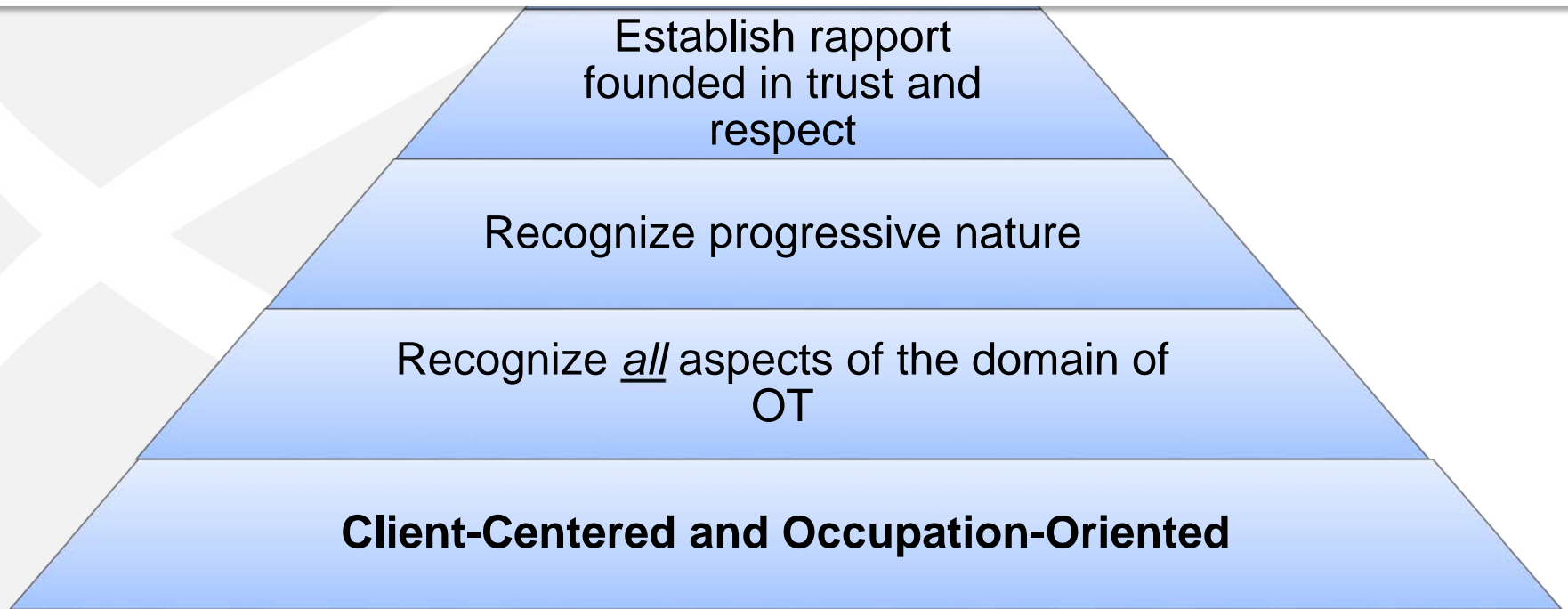
Knewstep-Watkins

# Building Blocks for OT Intervention



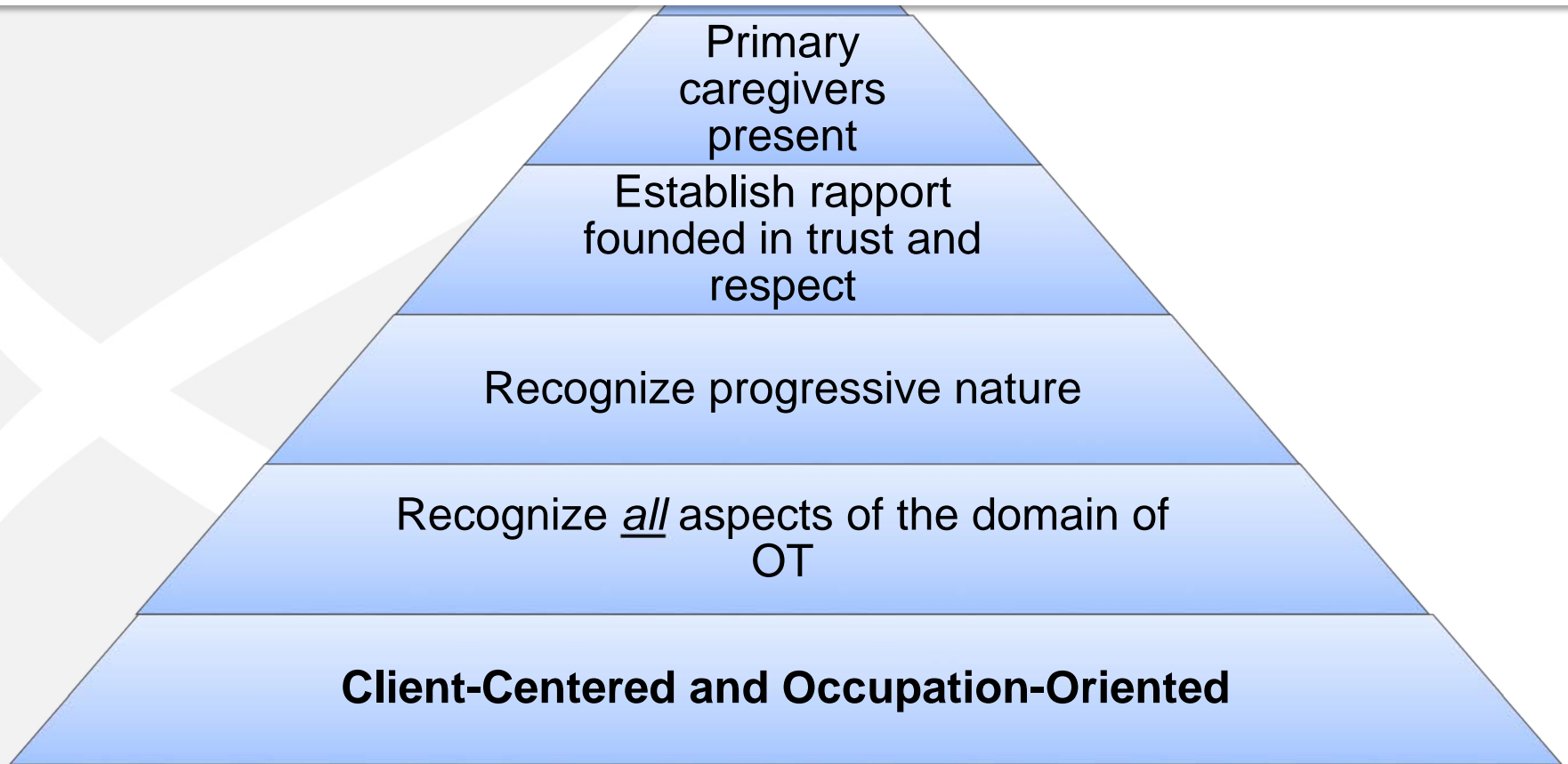
Knewstep-Watkins

# Building Blocks for OT Intervention



Knewstep-Watkins

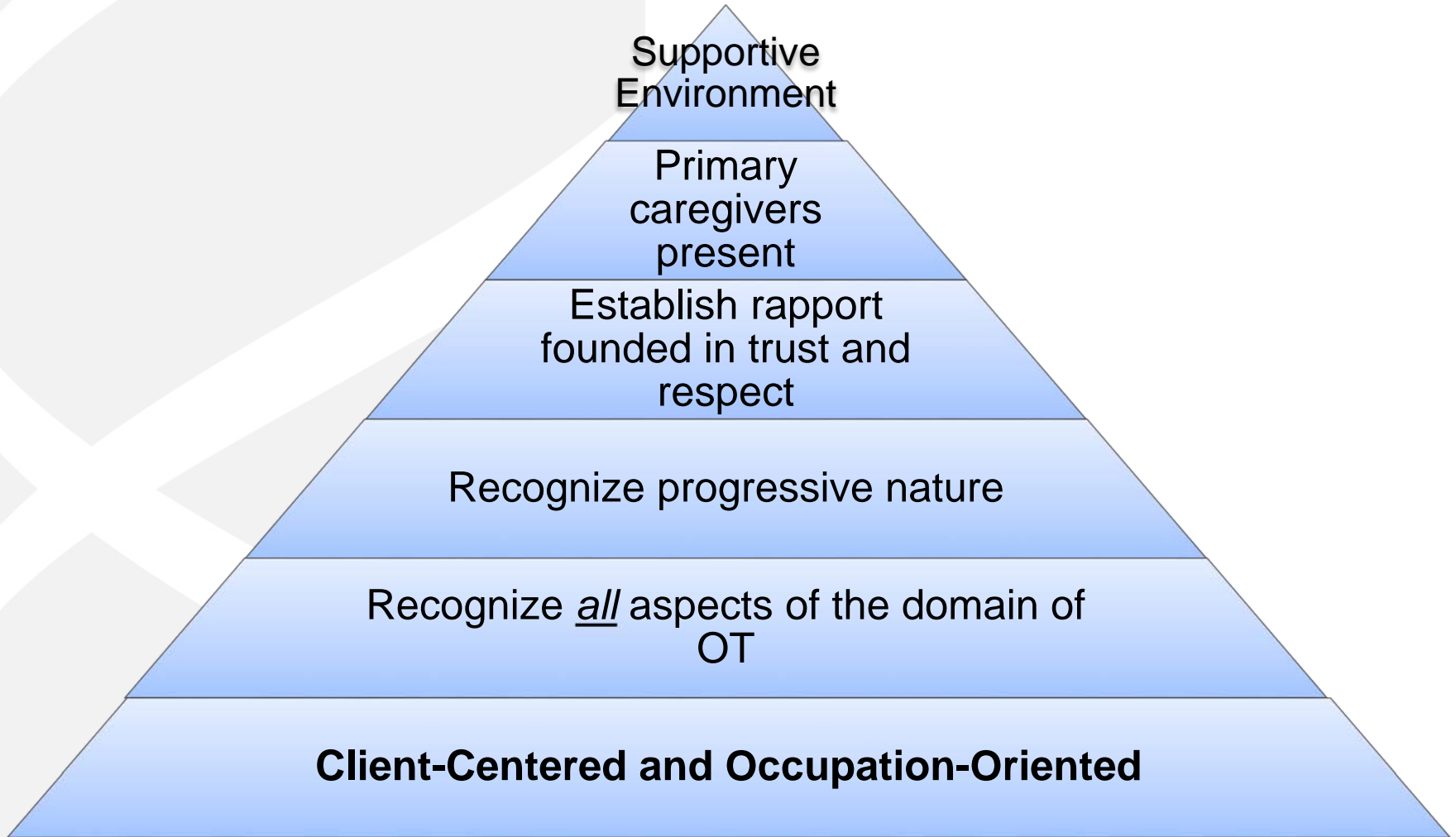
# Building Blocks for OT Intervention



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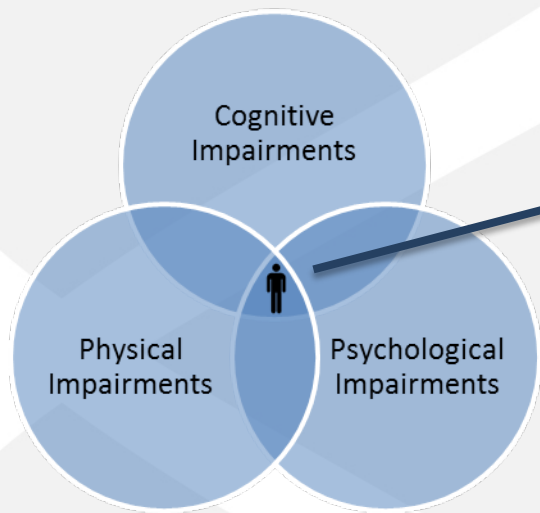


# Building Blocks for OT Intervention



Knewstep-Watkins

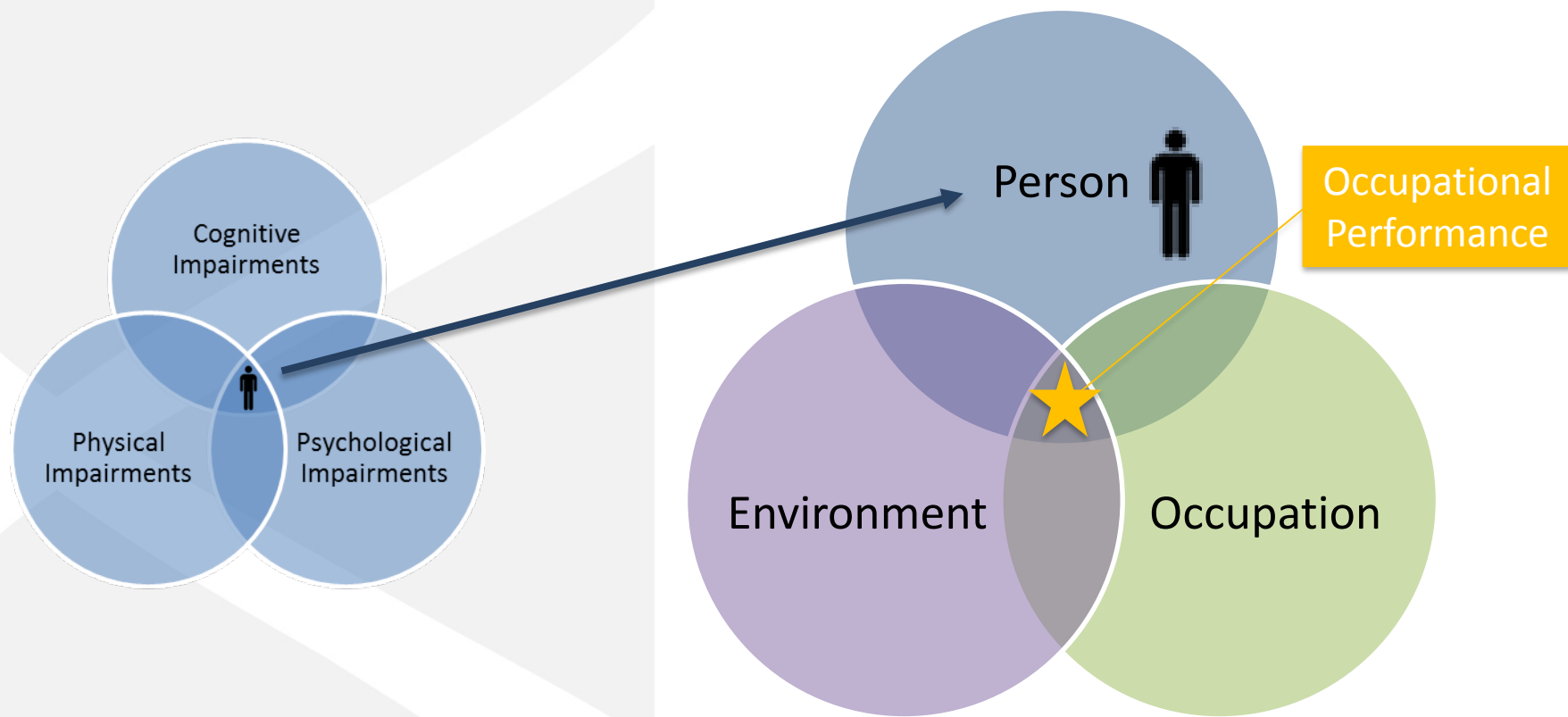
# OT for the Person with HD



OT Model of Practice?

Knewstep-Watkins

# OT: The Person-Environment-Occupation Model



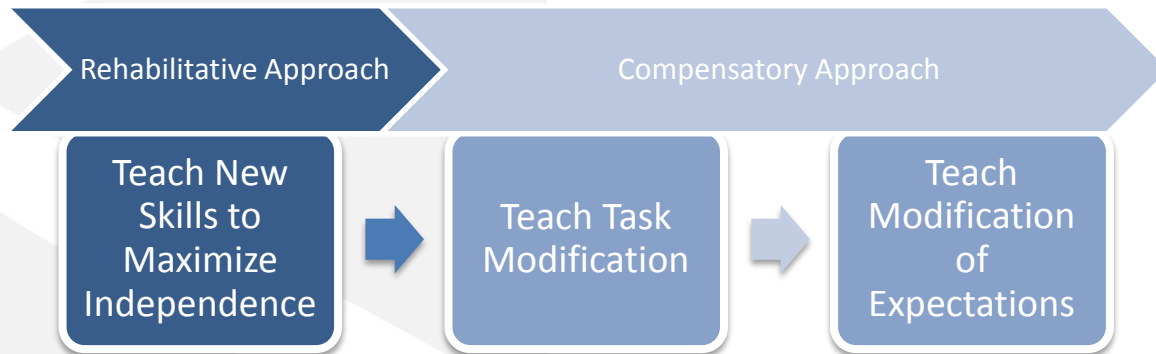
Knewstep-Watkins

# *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



Difficulty at Work  
Cognitive?  
Physical?  
Behavior?



Loss of Job



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 HD SA COE or through [HD SA](#)*

# EMPLOYMENT

**1<sup>st</sup>: Collaborate with patient, family, and social worker**

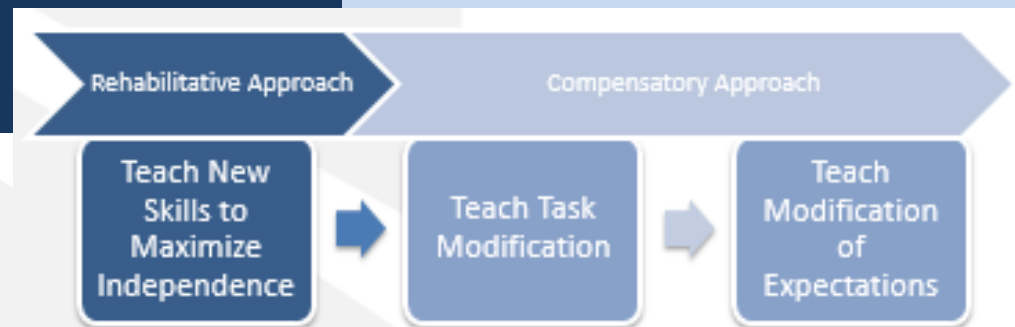
**2<sup>nd</sup>: Determine Rehabilitative vs. Compensatory**

## Rehabilitative Approach

- Consider Vocational Rehabilitation (state program)

## Compensatory Approach

- Most commonly used
- General recommendations

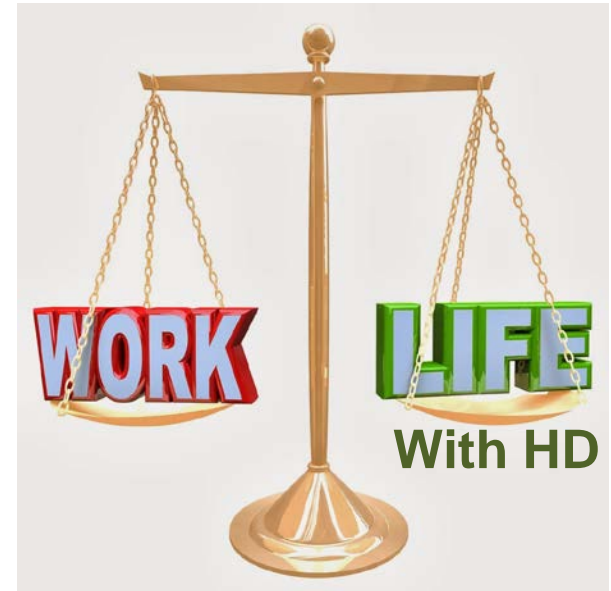




# EMPLOYMENT:

## General Compensatory Recommendations

- Reduce distractions
- Adjust work duties
- Adjust work hours
- Reduce commute
- Change jobs
- Additional Considerations:
  - Types of Harm
  - Burden of Stress



# Volunteering

**The just-right-fit may be volunteering!**

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

Examples:

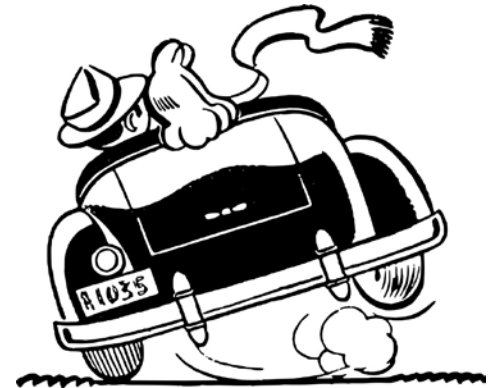
- Volunteering at a SPCA
- Volunteering for a church or civic organization
- Being involved in your local HDSA Chapter



# DRIVING

## Safe driving:

- Complex task that requires reliable:
  - ✓ cognitive abilities
  - ✓ motor abilities
  - ✓ **visual processing**
  - ✓ **control of one's behavior**



*I have spread my dreams under your feet;  
Tread softly because you tread on my dreams.*

*W.B. Yeats*

# 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?

# DRIVING

Mostly common approaches to retire from driving:



## 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](#))

# PUBLIC TRANSPORTATION



# LEISURE and SOCIAL PARTICIPATION

**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?**
- ✓ **2<sup>nd</sup>: Collaborative problem solving and sharing among OT, person with HD, and their family/friends.**
- ✓ **3<sup>rd</sup>: 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)

# LEISURE

## In Joe's words:

*Always an athlete, Joe remains active – staving off the physical effects of 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

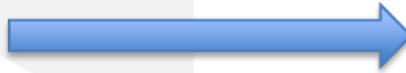
## Outdoorsman

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



## Gardeners

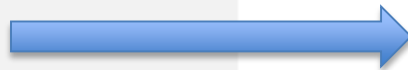
- Try a container and/or patio garden.



# 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](#)



### HD Workout Tips

#### Shana Verstegen's HD Workout Tips

In collaboration with the Huntington's Disease Society of America, Shana Verstegen has volunteered her time & expertise to provide valuable tips to help HD families live a healthier lifestyle.

HDSA provides Shana's workout methods as a courtesy to the HD community. HDSA, however, does not have fitness expertise. Please use caution when attempting these or any training methods or exercises. The videos are for motivation and teaching purposes only and do not replace the advice and personal attention of a fitness expert. See a doctor before starting any fitness program.



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[More Info](#)



THE BENEFITS OF EXERCISE FOR PEOPLE WITH  
HUNTINGTON DISEASE

# LEISURE: Exercise

## Tai Chi

### **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!***



# LEISURE and SOCIAL PARTICIPATION

## *Daisy's Mom Makes Impressive Sculptures*



“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.”

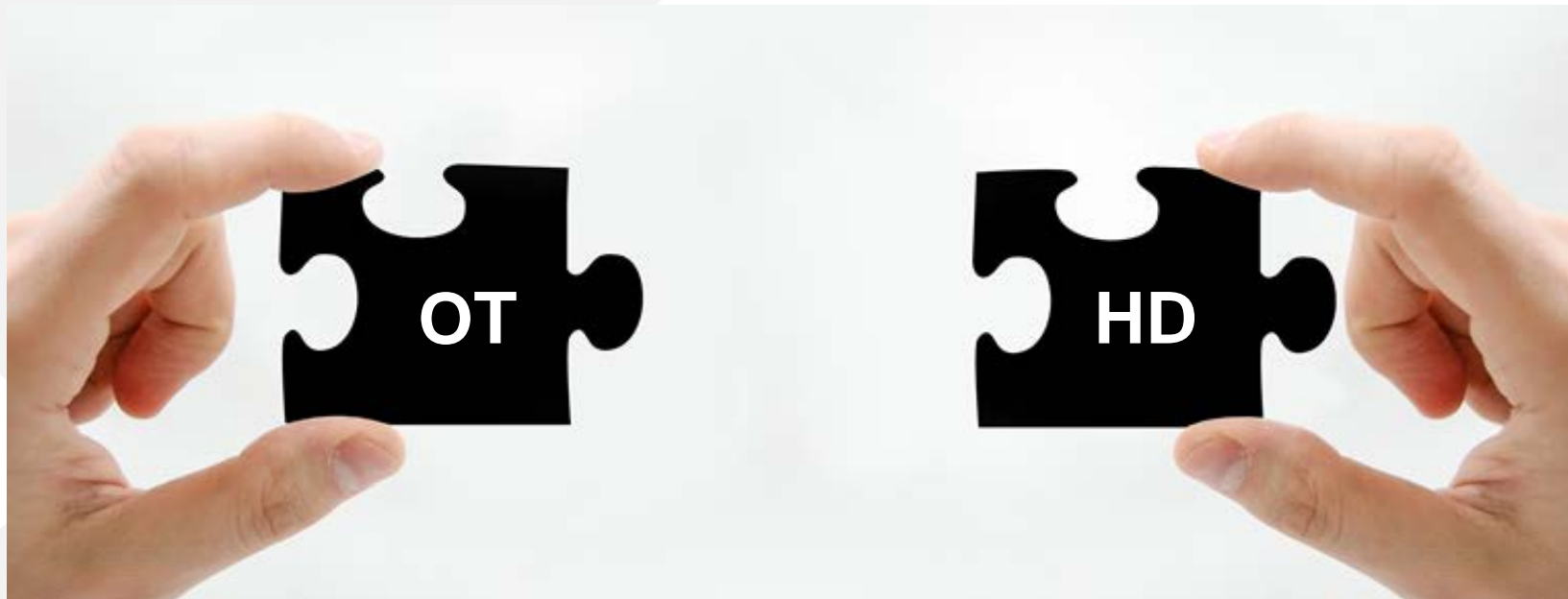
# LEISURE and SOCIAL PARTICIPATION: VACATIONING



***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**

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# 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)