Caregiving for People with Huntington’s Disease

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Learning Objectives

- Identify motor & nonmotor symptoms of HD
- Identify the stages of HD
- Identify strategies to educate and cue people as they progress through each stage of HD
- Identify appropriate assistive devices for people with HD
- Identify appropriate safe patient handling equipment to assist with a person’s functional mobility
- Identify techniques to assist people with functional mobility based on their deficits associated with HD
Presenter Disclosures

I have NO personal financial relationships with commercial interests relevant to this presentation.
Huntington’s Disease Facts

- **Incidence:** 5-10 per 100,000 (>15,000 per year) in U.S.
- **Prevalence:** At least 30,000 people in U.S.
- **150,000-200,000 AT RISK** due to having a 1st degree relative
- **Onset:** 30s-40s, but juvenile- (<20) & late- (>60) onset occur
- **Genetic testing may only suggest % risk of developing HD symptoms, but will not determine course or severity of disease.**
- Diagnosis is based on hx & clinical exam of motor features
HD Motor symptoms

Chorea

- Involuntary, irregular, nonrhythmic, abrupt, rapid, nonsustained movements of parts of the body.
- Can affect a body part, a limb, or the whole body.
- Unpredictable (unlike a tremor).
- Can be partially suppressed with concentration.
- Worsens with stress, anxiety, or when distracted.
- **Motor impersistence**: Inability to sustain isometric muscle contraction (poor motor control, not weakness)
Other Motor abnormalities

- **Dystonia**: Repeated twisting postures of body due to opposing muscle contractions
- **Akathisia**: Inner restlessness/anxiety that drives movement
- **Myoclonus**: Sudden, brief shock-like jerks
- **Tremors**: Oscillatory, rhythmic movements
- **Tics**: Quick, stereotyped, repetitive, but nonrhythmic movements or vocalizations
- **Parkinsonism**: Rigidity & slowness of movement that usually occurs in late-stage disease
HD Cognitive symptoms

- **Bradyphrenia**: Slowness of thinking
- **Executive Dysfunction**: Difficulty with attention, organizing, planning, multitasking.
- **Cognitive inflexibility**: Difficulty changing one’s mind
- **Memory loss** with impaired learning
- **Limited insight** and poor emotional recognition
- **Visuospatial dysfunction**: Poor depth perception/body awareness
- **Anosognosia**: *Unawareness* of HD that is not denial. Poor insight into impairments, functional limitations, and behavior issues.
HD Psychiatric symptoms

- Depression - hopelessness may increase risk for suicidal ideation
- Anxiety - often leading to panic attacks
- Obsessions - often paired with compulsive behavior
- Mania - insomnia, distractibility, irritability, impulsive risk-taking
- Delusions - often of grandiosity
- Hallucinations
HD Behavioral Manifestations (Dysexecutive Syndrome)

- **Irritability** - easily agitated
- **Disinhibition** - disregard of accepted social norms
- **Impulsivity** - acting without consideration of consequences
- **Perseveration** - fixation on idea or action/behavior
- **Apathy** - loss of motivation/initiation
- **Ambivalence** - inability to make choices or opinions
Clincial Course of HD

Pre-Manifest Period
- 10-15 years before onset of HD
- No symptoms of disease

Prodromal Period
- Subtle motor, cognitive, and behavioral changes.
- Family or person with eventual diagnosis of HD may realize changes only in retrospect.

Manifest Period
1. Still independent
2. Reduced job capacity, may still drive, but needs help with IADLs
3. Unable to work, needs supervision, impaired ADLs
4. Needs 24/7 support and help with all ADLs
5. Dependent for total care
Progression of symptoms and disability in a typical person with Huntington’s Disease

Prodromal Period

Pre-Manifest Period

(Nance et al, 2011)
Rehabilitation Along the Continuum of Care

Pre-Manifest-Early Stage-1

Neuroprotection Wellness
- Forced Exercise Paradigm
- Intensive Aerobic Exercise
- Prevention

Middle stage-2

Neoplasticity Motor Learning
- Gait Training
- Dual-Task Practice
- Aerobic Exercise
- Task-Specific Training
- Postural Stability
- Posture
- Flexibility
- Strengthening

Middle stage-3

Neoplasticity Wellness
- Gait
- Dual-Task Practice
- Aerobic Exercise
- Balance
- Posture
- Flexibility
- Strengthening

Late stages 4-5

Compensation Adaptive Rehab
- Home Safety
- Fall Prevention
- Family Training
- Adaptive Equipment

Preventative

Restorative

Compensatory

Figure 17.1

Summary of the rehabilitation approach across the continuum

(Hernandez et al, 2015)
**Education for Pre-manifest & S&F Stage 1**

- Promote moderate-to-high intensity **cardio** (60-85% HR max) 3-5X/week for 30-60 minutes. May accumulate cardio in bouts of 10-minute intervals. **Inactivity facilitates degeneration!**

- **Compliance matters**, so make sure the person enjoys activity!

- Stretching only tight muscle groups, if needed, 30-60 seconds.

- Strengthening (especially large muscle groups) 2-3 times per week performing 1-3 sets of 8-12 reps per group.

- Balance Training 2-3 times per week (Yoga, Pilates, Tai Chi)

**Take home message from UCD HD Center:**

More active people with neurodegenerative disease always do better at maintaining their functional independence longer!
Why promote cardio?

- We all know it's good for the heart, lungs, metabolism, etc...
- Intensity & duration of cardio correlates with increased levels of Brain-Derived Neurotrophic Factor (BNDF)

What?

- **BNDF** is a protein that supports differentiation, maturation, and survival of neurons in the nervous system and shows a neuroprotective effect under adverse conditions, such as cerebral ischemia, hypoglycemia, and neurotoxicity.
- BDNF also stimulates and controls growth of new neurons from neural stem cells (neurogenesis).
- Brain-derived neurotrophic factor is *probably* neuroprotective in PD & HD!!
Exercise in HD
Visit HDSA.ORG for links to exercise videos
Middle stage of HD (S & F Stages 2-3)

- **Dysarthria** - Slurring of words due to facial weakness
- **Dysprosody** - Impaired ability to control intonation, intensity, vocal quality, and rhythm of speech.
- **Dysphagia** - Difficulty swallowing may begin. Important caregivers recognize signs of choking and Heimlich maneuver!
- Cognitive impairment affecting attention, ability to multi-task, judgment, motor planning, and spatial perception.
- Psychiatric and behavioral problems may evolve.
- Worsening of gait, balance, coordination, and endurance.
- Stress and anxiety exacerbate chorea and irritability.
Middle stage of HD (S & F Stages 2-3)

- Create a routine schedule with planned routine variation (to increase ease of transition for errands/appointments).
- Allow extra time for ADLs and other tasks; avoid time pressure.
- Structure the environment, remove tripping hazards, pad furniture, consider carpeting floors (but avoid throw rugs).
- Encourage use of assistive devices and adaptive equipment early on to form habits before ability to learn declines.
- Engage the person in errorless learning as cognition declines.
- Avoid multi-tasking & be cautious in distracting environments.
- Keep instructions simple, ask closed-ended questions (or limited choice options), and consider communication board.
- Do not infantilize the person or talk down to them.
Assistive Devices & Environment Mods

How to Prevent Falls

30% of falls can be prevented. Here's how:

Make Your Home a Safer Place:

- Install handrails on staircases
- Pick up clothes
- Clear away loose cords
- Use non-slip mats in shower
- Add more lighting
- Declutter Kitchen
Guarding during mobility/walking during S&F Stages 3-4 (mid-late)

- Unlike PD, predicting direction of falls is difficult.
- Chorea is not predictable, so guarding is like dancing.
- Increased risk for agitation if over-guarding or restricting movement.
- Strongly recommend 4WW & appropriate footwear!
- Use a gait belt, but move at the patient’s pace
- Sometimes keeping lots of padded furniture around to enable “furniture cruising” is acceptable if patient refuses to use AD.
- Consider carpeting floor and use bed, chair, or motion alarms.
Ideas for reducing falls

FIGURE 7-34 Anti-tipping device.
Adaptive Bathroom Equipment

[Image of adaptive bathroom equipment: a toilet with support bars and an adjustable chair with red arrows indicating movement and adjustment features.]
Handling Dysexecutive Syndrome and Outbursts

- Schedules, routines, and gentle reminders (AV record?)
- Always remain calm—be pleasant, but firm
- De-escalate with norm of reciprocity
- Avoid over-correction, confrontations, and ultimatums.
- Focus on shared outcome goals and what the patient can do, rather than what they can’t. Re-direct them.
- Be aware of triggers and signals that precede outbursts.
- Be aware of your own body language, don’t be threatening.
- Leave if situation escalates & threatens combative behavior.
Late Stage HD (S&F Stages 4-5)

- Person will need more support if ambulatory.
- Personal protective padding (helmets) can be encouraged.
- If person needs help to stand, sometimes having them hug themselves will diminish choreic limb movements.
- Consider safe patient handling devices—Standing aid or Hoyer lift.
- Put mattress on floor if patient tends to vault OOB onto floor due to poor force modulation, but assumes floor recovery skills.
- Consider padded bed rails. (Posey bed may increase agitation.)

Equipment should optimize patient’s independent movement, but balance safety of the patient and the needs of the caregiver.
Trouble with walking & falls in HD

Gait Belt w/ handles

Padding
Protection from furniture...
Caregiver techniques for Stages 4-5

- Dystonia/Rigidity
  - Flex great toe while pushing on hamstring to help relax leg
  - Segmental rolling
  - Use appropriate equipment to help to safely handle the person
  - Caregiver training, positioning, patient handling, hygiene, etc.

Flex the big toe! Push behind knee!
Draw sheets & Slip sheets
Sit->Sidelying->Supine: Dependent
Supine->Sit: “Flat Spin” (best with 2 people)
Preventing falls from bed: Block knees and prevent posterior lean!
Blocking Trunk, Hip, & Knee/Foot
For standing or transferring

Face the patient from the side.

Front arm supports axilla.

Back arm pulls glute into extension.

Front leg blocks knee into extension.

Front foot keeps patient from sliding.

You must squat and pull, not lift.

Use momentum to rock the person forward “nose over toes”.

Stand on the count of three...
Verbal cues to help a person stand up

IF I wanted to stand up, Am I using optimal body Mechanics in this photo?

Standing Mechanics
1. Scoot forward
2. Bend knees >90 degrees
3. Plant feet shoulder width
4. Elevate surface?
5. Lean forward: “Nose over toes!”
6. Push off with 1-2 arms
7. Use momentum, if needed
Assisted squat-pivot transfer with 3 points of contact: Knee, Hip, Trunk
Always have draw sheets in place!

Example of a dependent squat-pivot transfer technique.

Patient shoulder to your chest for trunk control.
You control hips with sling created by sheet.
Knee block to prevent buckle or foot slide.
In this position I could swing her easily to left.
Note: You block and sit back, don’t lift patient!
Equipment
Draw sheets give you options!

Gives you better hip control for 2-person assisted standing.

Enables you to reposition patients easily. See next slide for scooting patient in chair.
Scooting dependent patient backward in a wheelchair

1. Lean patient forward.
2. Use arm to prevent forward fall.
3. Grab draw sheet under upper thigh.
4. Grab draw sheet under ischium.
5. Pull up on draw sheet under ischium.
6. Patient will slide back.
Reducing anterior slide from chair
Enlist the help of professionals to assist you in determining how best to care for people with HD.

Device prescription, Caregiver training, Floor recovery training, etc.
For more information on HD

- Huntington’s Disease Society of America: https://hdsa.org/


Questions?

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Photo credits, Product URLs, and References on following slides


Photo credits and product URLs

- [https://www.hdis.com/gait-transfer-belt?gclid=EAIaIQobChMlk-im5_145AIVtx-tBh0W9gfUEAQYBCABEgL3vD_BwE#155=1561?cid=199755&dest=](Gait belt with handles)
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- [https://www.amazon.com/Kxtffeect-Premium-Furniture-Proofing-Protector/](https://www.amazon.com/Kxtffeect-Premium-Furniture-Proofing-Protector/) (Furniture pads)
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- [https://www.vitalitymedical.com/lumex-stand-assist-patient-transport-graham-field-lf1600.html?network=g&device=c&keyword=&campaign=1031885966&adgroup=pla-341242296360&gclid=EAIaIQobChMIenwn_X75AlVWyCtBh3NkADDEAAYASABEgJCAfD_BwE](https://www.vitalitymedical.com/lumex-stand-assist-patient-transport-graham-field-lf1600.html?network=g&device=c&keyword=&campaign=1031885966&adgroup=pla-341242296360&gclid=EAIaIQobChMIenwn_X75AlVWyCtBh3NkADDEAAYASABEgJCAfD_BwE) (Stand assist aid)
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