



A Roadmap for HD - What comes next?



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Presenter Disclosures

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

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Our Agenda

- Introduction to HD
- Definition of stages
- Description of scales used to identify stages
- Discuss findings in early HD from clinical trials
- Discuss each stage with a focus on the late stage and treatment strategies



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Our First Quiz – TEXAS (True or False)



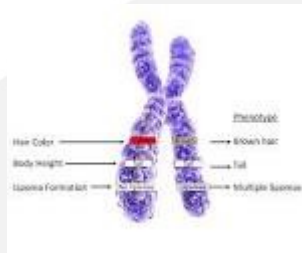
- 1) All Texans ride horses to work or school.
- 2) Texas is popularly known as The Lone Star State.
- 3) The first word spoken from the moon on July 20, 1969 was Houston.

The Big Picture - HD

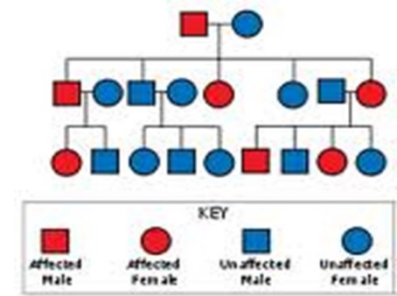


- Huntington's disease (HD) is a SLOWLY progressive neurodegenerative disease.
- HD encompasses a triad of motor, cognitive and psychiatric symptoms.
- We have effective symptomatic therapies (medical and nonmedical) for all stages of HD **-treatable**.
- The average duration of survival after a clinical diagnosis is 15-20 years.
- Everyone is on their own path....
- You are not alone!





Genetics of HD

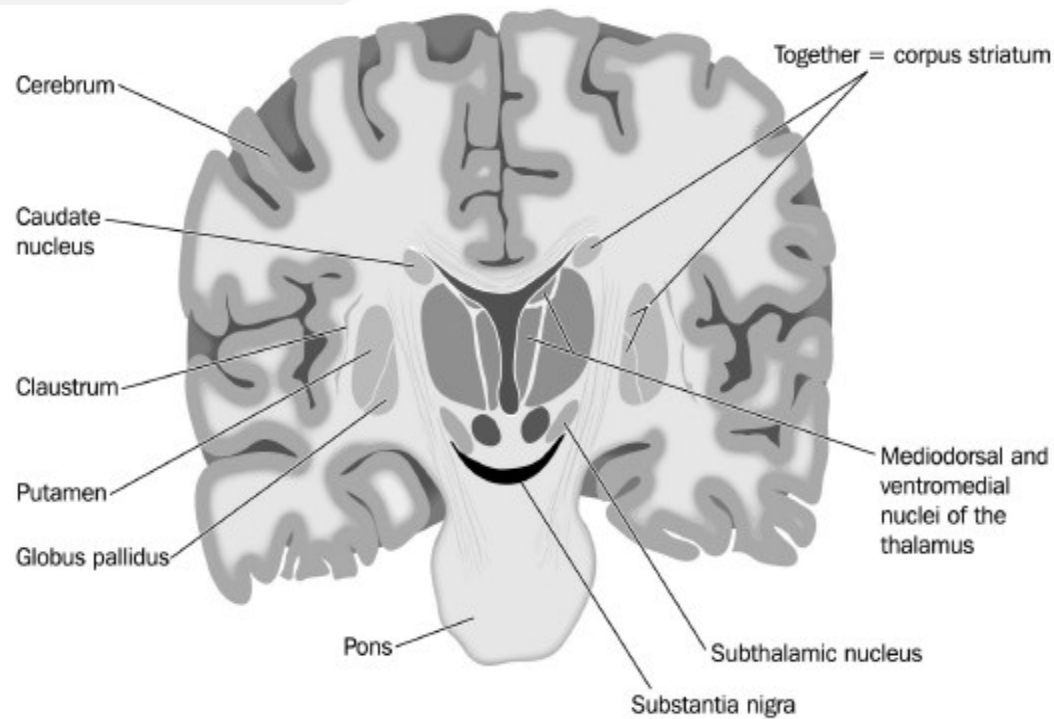


- Genetics: Autosomal dominant pattern of inheritance –
 - Affects males and females equally
- Each child of an affected parent has a 50% chance of inheriting the HD gene.
- HD is caused by trinucleotide repeat expansion in the huntingtin (HTT) gene (aka HD and IT15) on chromosome 4.
- All humans have 2 copies of the HTT gene.
 - The HTT gene encodes for the protein huntingtin.
 - When the trinucleotide repeat exceeds the normal range, the protein produced is abnormal (mutant Huntingtin protein mHtt).
 - The abnormal protein is thought to be toxic to neurons.

HD Genetics

- HD is caused by a mutation in the Huntingtin gene
 - Type of mutation: —CAG repeat expansion
 - 10-35 CAG is normal
 - > 35 is not normal and can cause HD
- •Everyone with HD has a CAG repeat expansion in the HD gene
- •CAG repeat expansions between 36-39 may not lead to symptoms within a normal lifespan

Coronal View of Important Structures



A bit of history...

- 1842 - Charles Oscar Waters wrote a letter published in the first edition of Robley Dunglison's *Practice of Medicine*.
 - Described “a form of chorea vulgarly called magrums”.
- 1872 - First thorough description of the disease was by George Huntington
- "Of its hereditary nature. When either or both the parents have shown manifestations of the disease ..., one or more of the offspring almost invariably suffer from the disease ... But if by any chance these children go through life without it, the thread is broken and the grandchildren and great-grandchildren of the original shakers may rest assured that they are free from the disease."

Huntington G (1872). ["On Chorea"](#). *Medical and Surgical Reporter of Philadelphia* (The Hague: Nijhoff) **26** (15): 317–321.

THE
MEDICAL AND SURGICAL REPORTER.

No. 789.]

PHILADELPHIA, APRIL 13, 1872.

[Vol. XXVI.—No. 15.]

ORIGINAL DEPARTMENT.

Communications.

ON CHOREA.

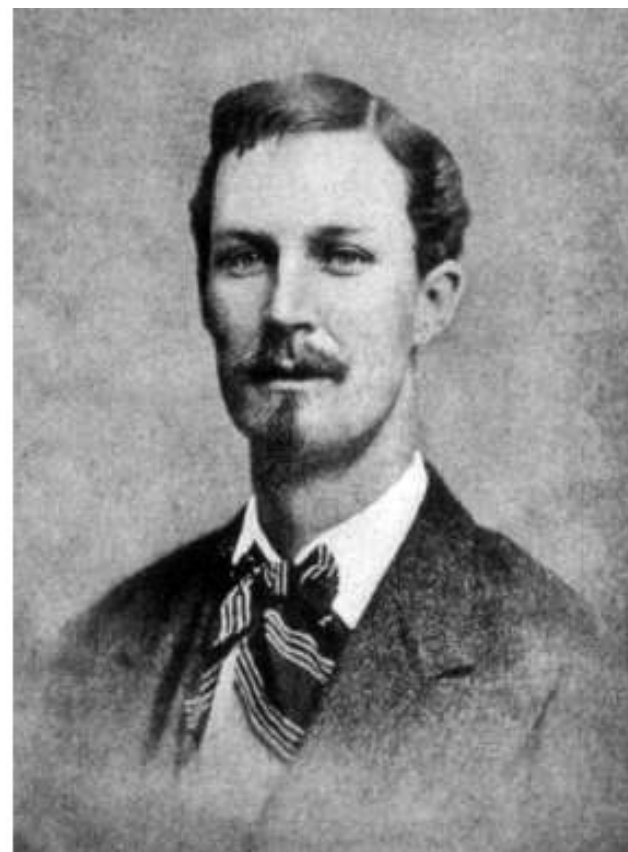
By GEORGE HUNTINGTON, M. D.,
Of Pomeroy, Ohio.

Essay read before the Meigs and Mason Academy of Medicine at Middleport, Ohio, February 15, 1872

Chorea is essentially a disease of the nervous system. The name "chorea" is given to the disease on account of the *dancing* propensities of those who are affected by it, and it is a very appropriate designation. The disease, as it is commonly seen, is by no means a dangerous or serious affection, however distressing it may be to the one suffering from it, or to his friends. Its most marked and char-

The upper extremities may be the first affected, or both simultaneously. All the voluntary muscles are liable to be affected, those of the face rarely being exempted.

If the patient attempt to protrude the tongue it is accomplished with a great deal of difficulty and uncertainty. The hands are kept rolling—first the palms upward, and then the backs. The shoulders are shrugged, and the feet and legs kept in perpetual motion; the toes are turned in, and then everted; one foot is thrown across the other, and then suddenly withdrawn, and, in short, every conceivable attitude and expression is assumed, and so varied and irregular are the motions gone through with, that a complete description of



Huntington's Disease
Society of America

Let's start at the very beginning...

A few definitions



- HD At-Risk - Individuals who have a biological parent with a diagnosis of HD who have not been tested are considered at 50% risk for HD.
- For persons to be considered at-risk, no signs or symptoms of HD are present.
- HD Expansion Carrier – Individuals who have undergone a predictive genetic test for the HD-causing CAG repeat length and have an expansion greater than 35 repeats. They have the gene for the disease, but they show no current signs or symptoms of HD. These individuals are preprodromal.

Definitions (cont.)

- HD Prodrome – at-risk persons who are years from the appearance of the motor symptoms
 - Observational studies on the earliest stages of HD have revealed that the some cognitive and behavioral symptoms indicating the presence of a disease process prior to the development of the full clinical syndrome and may negatively impact function and quality of life.
- The prodrome may appear up to 15 years before the onset of motor symptoms
- Manifest HD – patients with expanded CAG repeats that are clearly symptomatic

Stages of Manifest HD

- HD can be divided functionally into 3 stages –
“real world staging”

- 5 stage scale used clinically based on TFC

Early – individuals continue to drive, manage their finances and work independently

Middle – individuals may lose some independence (might lose the ability to drive or manage their finances), but will continue to perform their activities of daily living (ADLs) independently.

Late – individuals require assistance with ADLs

Unified Huntington's Disease Rating Scale (UHDRS)

- A validated rating scale to assess the clinical features of HD
 - Developed by the HSG
 - Published in 1996
- Motor
- Cognitive
- Behavioral
- Independence
- Functional assessment
- Total functional capacity

UHDRS – Motor/Chorea subscale assessment

Unified Huntington's Disease Rating Scale Motor Assessment Chorea Scale		
Body Region	Severity	
Face	0	Absent
Bucco-oral-lingual	1	Slight/intermittent
Trunk	2	Mild/common or moderate/intermittent
Right upper extremity	3	Moderate/common
Left upper extremity	4	Marked/prolonged
Right lower extremity	Total score: Sum of scores for each body region Range = 0 - 28	
Left lower extremity		

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Shoulson-Fahn Total Functional Capacity Rating Scale

Total Functional Capacity Rating Scale ¹ (Source: UHDRS ²)		
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
TOTAL	Range 0 - 13	

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

1 Shoulson I, Kurlan R, Rubin AJ et al. Assessment of functional capacity in neurodegenerative movement disorders: Huntington's disease as a prototype, in Munsat TL (ed): Quantification of Neurological Deficit. Boston: Butterworth, 1989, pp 271-283.

2 The Huntington Study Group. Unified Huntington's Disease Rating Scale: reliability and consistency. Mov Disord 1996; 11:136-142.

Shoulson and Fahn Staging Scale¹

Stages based on TFC

- HD Diagnosed STAGE I (0–8 years since motor diagnosis):
- Marginal engagement in occupation, having part-time voluntary or salaried employment potential AND maintains typical pre-disease levels of independence in all other basic functions, such as financial management, domestic responsibilities, and activities of daily living (eating, dressing, bathing, etc.);
- OR performs satisfactorily in typical salaried employment (perhaps at a lower level) and requires slight assistance in only one basic function: finances, domestic chores, or activities of daily living.
- Progression on this scale is about 1 point/year for both Stages I and II (Marder et al., 2000).

Stages based of the TFC

- HD Diagnosed STAGE II (3-13 years since motor diagnosis): Typically unable to work, requiring only slight assistance in all basic functions: finances, domestic, daily activities; OR unable to work and requiring different levels of assistance with basic functions (some are still handled independently).
- Progression on this scale is about 1 point/year for both Stages I and II.
- HD Diagnosed STAGE III (5-16 years since motor diagnosis): Unable to engage in employment AND requires major assistance in most basic functions: financial affairs, domestic responsibilities, and activities of daily living.
- Annual progression on the TFC is .38 point/year for this stage.

Stages based on TFC

- HD Diagnosed STAGE IV (9-21 years since motor diagnosis): Requires major assistance in financial affairs, domestic responsibilities, and most activities of daily living. For example, comprehension of the nature and purpose of procedures may be intact, but major assistance is required to act on them.
- Care may be provided at home but needs may be better provided at an extended care facility.
- Annual decline on the TFC is .06 point/year for persons in this stage.
- HD Diagnosed STAGE V (11-26 years since motor diagnosis): Requires major assistance in financial affairs, domestic responsibilities, and all activities of daily living. Full-time skilled nursing care is required.

Earliest motor symptoms in HD Gene Carriers

- Individuals who carry the gene might notice the following:
 - Dropping objects more often
 - Fine hand coordination is slightly impaired
 - A bit more “fidgety”
 - Decreased dexterity while playing a musical instrument
 - Decreased sense of rhythm
 - Tripping more easily on uneven surfaces

Early Motor Symptoms in HD Gene Carriers

- On exam, the physician might note:
 - Subtle irregularity (speed and amplitude) in the rhythm of finger taps
 - Subtle irregularity (in speed, timing and amplitude) with rapid alternating movements
 - Subtle gait difficulty – tandem gait impaired
 - Subtle change with eye movements
 - Slowing of saccade velocity and initiation

Measurement of motor function in Pre-HD Individuals

- Objective measurements are important:
 - Increased subjective complaints of incoordination in an HD-gene carrier may just be the tendency for an HD-gene carrier to perceive any clumsiness as the first indication of disease appearance.
- Several large observational studies were conducted in HD gene carriers who were near or far from clinical diagnosis.
- PREDICT-HD and TRACK-HD were large multicenter prospective observational studies that examined clinical and biological findings of disease progression in individuals with premanifest HD (preHD) and early-stage HD.

Motor Abnormalities in Pre-manifest HD (PREDICT-HD Study)

- Designed to identify clinical and biological markers in premanifest individuals who have undergone predictive genetic testing.
- Baseline motor data between gene-expansion carriers (cases) and non gene-expansion carriers (controls) were compared.
- Cases were categorized as near (<9 yrs), mid (9-15yrs) or far (>15 yrs) from diagnosis using a CAG-based formula (far from estimated diagnosis.
- Participants were recruited from 30 sites in the United States, Canada, Australia and Europe

Mov Disord. 2009 September 15; 24(12): 1763–1772.

Results from PREDICT-HD (Motor Function)

- Pre-HD cases with closer estimated proximity to diagnosis had worse total motor scores and worse scores on the motor domains than individuals further from estimated diagnosis.
- Most apparent for total motor scores, chorea domain, bradykinesia domain and the oculomotor domain.
- These findings are consistent with other reports that suggest chorea and quantitative measures of eye movements may be sensitive markers in pre-HD individuals.
- Finger tapping and tandem gait performance, saccade velocity and chorea scores were inversely associated with striatal volume.

Mov Disord. 2009 September 15; 24(12): 1763–1772.

PREDICT-HD Motor Summary

- Subtle motor abnormalities were present in pre-HD individuals.
- These motor abnormalities were associated with closer proximity to estimated disease diagnosis and greater striatal atrophy.
- These findings suggest that the UHDRS motor examination may be a useful outcome measure in clinical trials aimed at delaying diagnosis.

TRACK-HD Study

- The purpose of this observational study was to answer the critical
- question:
 - When should drugs that might delay onset of HD be started?
 - Subjects were recruited from Canada, France, UK, Netherlands
- Participants were thoroughly examined every year for 3 years
- Dozens of measurements were made on each subject that included:
 - Neuroimaging (MRI of brain)
 - Motor symptoms (including eye movement tracking)
 - Intellectual (cognitive) function
 - Emotional well being

www.thelancet.com/neurology Vol 12 July 2013

TRACK-HD Study

- Subjects without symptoms of HD (but gene carriers) were divided into two groups: those who are estimated to be close to or far from disease onset:
- Predicting how close subjects were to onset was based on a mathematical calculation.
- A group of subjects in the early stages of HD and a control population that didn't carry the gene were also studied.
- 366 subjects were enrolled and 298 were able to complete the 36 month follow up (those that dropped out of the study were in the more advanced stages of disease).

Summary of TRACK - HD

- Early changes in motor control can be observed and measured in pre-manifest HD in carriers of the HD gene.
- Total motor scores measured in the clinical examination (UHDRS) are increased in pre-HD persons who are estimated to be near diagnosis.
- Quantitative measurements are much more sensitive than a clinical diagnosis.
 - Finger tapping (inter-tap interval duration and variability) was one of the best motor indicators of progression along the trajectory from pre-HD to clinically manifest HD.

Quiz # 2 - HD

True or False

- 1) Huntington's disease is a rapidly progressive disease.
- 2) Individuals with HD may experience cognitive, motor and behavioral symptoms.
- 3). The are effective available symptomatic therapies for patients with HD.

Early Stage

- Movements may be minimal.
- Individuals remain functional and independent.
- Changes in mood will likely occur such as:
 - Depression
 - Individuals may say they feel discouraged, sad, hopeless, unmotivated, or disinterested.
 - Suicidal ideation may occur.
 - Anxiety
 - Pts may report feeling nervous, more irritable, and report problems sleeping and/or concentrating.

Psychiatric Features of HD

- Behavioral changes can be most distressing to patients and caregivers but are TREATABLE.
- – Depression is common in HD, often precedes diagnosis and may be most prominent in the early stages.

Symptoms of Major Depressive Disorder (MDD) include:

- Depressed mood most of the day, nearly every day
- Decreased ability to find pleasure or interest in usual activities
- Significant decrease or increase in appetite or weight
- Fatigue or loss of energy nearly every day
- Changes in sleep – insomnia or hypersomnia
- Restlessness or feeling physically slower
- Decreased ability to think, concentrate, and make decisions
- Feelings of guilt or worthlessness
- Recurrent thoughts of death or committing suicide.

- Source: *Diagnostic and Statistical Manual of Mental Disorders, DSM-5*

Treatment Examples

Some Psychiatric Medications Commonly Used in Treating the Psychiatric Symptoms of HD

Note: There is only one medication that is specifically approved by the FDA for use in HD – Tetrabenazine, a chorea treatment. The following medications are suggestions based on the clinical experience of the author. Physicians should carefully review the pharmaceutical manufacturers' materials regarding dosage and potential side effects before prescribing any medication.

Antidepressants

(used for depression and sometimes for irritability and anxiety as well):

SSRIs:

- | | |
|---------------------------|------------------------|
| - Sertraline (Zoloft®) | - Paroxetine (Paxil®) |
| - Citalopram (Celexa®) | - Fluoxetine (Prozac®) |
| - Escitalopram (Lexapro®) | |

Others:

- | | |
|---------------------------|--------------------------|
| - Bupropion (Wellbutrin®) | - Venlafaxine (Effexor®) |
|---------------------------|--------------------------|

Neuroleptics

(used for psychosis and sometimes for irritability or for chorea suppression)

- Atypical Antipsychotics:

- | | |
|--------------------------|---------------------------|
| - Olanzapine (Zyprexa®) | - Ziprasidone (Geodon®) |
| - Quetiapine (Seroquel®) | - Aripiprazole (Abilify®) |

High Potency Antipsychotics (sometimes also used for chorea suppression) :

- | | |
|----------------------------|----------------------------|
| - Haloperidol (Haldol®) | - Risperidone (Risperdal®) |
| - Fluphenazine (Prolixin®) | |

Suicide and HD

- Suicide is more common in patients with HD than the general population.
- Suicide is preventable.
- Red flags - If an individual with HD withdraws or reports feeling hopeless or “wanting to give up.”
- Encourage the individual to talk about his or her feelings.
- Ask the person if they have a specific suicide plan – call your MD or a suicide prevention line.
- Remove any potential weapons, medications, sources of danger from the home.
- Don’t be afraid to ask your loved one about suicidal ideations if you are concerned.

Behavioral Changes

- Obsessive and Compulsive Behaviors
 - Obsessions: involuntary, uncontrollable thoughts, or impulses that occur repeatedly.
 - Compulsions: behaviors or rituals that one acts out repeatedly.
 - Compulsive hand washing/ worried about contamination
 - Stuck to a ritual
 - May become fixated on a past episode of feeling wronged.
 - Difficulty with changes to routine
 - Perseveration: redundant repetition
 - Work towards redirecting the individual
 - Medications are available if quality of life is negatively impacted.

Early Stage

- Might notice subtle changes in cognition such as:
- Difficulty with sequencing or planning, decrease in the ability to focus, concentrate.
- Patients might seem unaware of these changes.
- Tips for caregivers...
- Individuals with HD might have difficulty with awareness of symptoms or “organic denial”.
- Recognize that the HD patient might not ever “accept” the disease
 - Insight might always be limited and that is okay.
- Allow the person with HD to talk about the challenges they are experiencing on a day to day basis.

Early Stage and Treatment

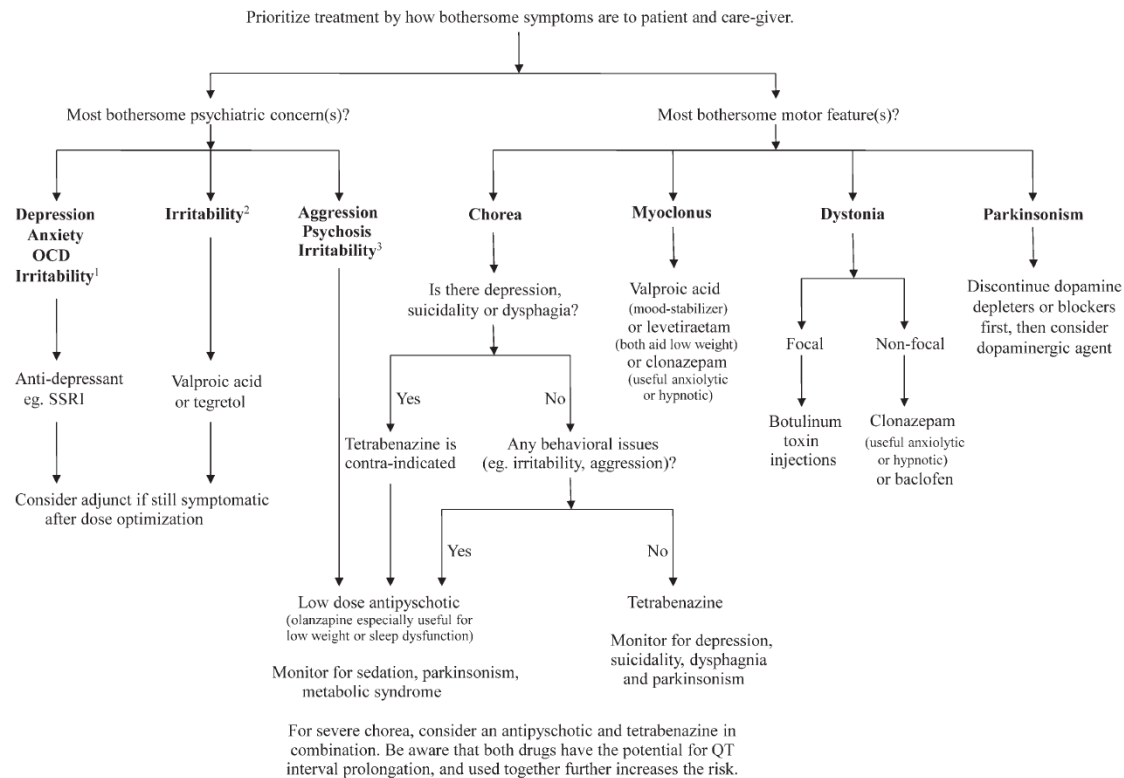
- Exercise - beneficial and therapeutic ? Neuromodulatory?
- Multiple clinical trials in PD support the efficacy of exercise
- No perfect regimen identified – stay active!
- Physical therapy (neurological therapy if available)
 - Optimize gait, balance and posture
 - Medications available for involuntary movements are available, may not be needed in early HD.
 - Discuss your concerns in detail with your physician.

Multi disciplinary Team for HD

Health Professional	Role in HD Care
Neurologist	Overall care coordination ("team leader"); neurological management
Psychiatrist	Overall care coordination ("team leader"); psychiatric management
Psychologist	Counseling about relationships, grief, chronic disease, symptom management, etc.; family support
Neuropsychologist	Neuropsychological assessment and counseling about cognitive changes
Geneticist or Genetic counselor	Genetic counseling, predictive testing, prenatal or preimplantation genetic testing
Social worker	Disability, financial planning, management of social crises, accessing community services, placement outside the home; Advance Directives; counseling (if licensed)
Physical therapy	Gait assessment and assistive devices; exercise program
Occupational therapy	Assessment of driving, home safety, and activities of daily living; equipment for seating, feeding, hygiene, etc.
Speech therapy	Speech assessment, alternative communication devices; dysphagia assessment and counseling
Dietician	Nutritional assessment and counseling
Nursing	Case management, telephone counseling, support
Research team	Counseling about research opportunities; enrollment in research
Chaplain	Family support, spiritual advice
Lay organization volunteer	Liaison with HD support groups, advocacy, other organization activities
Primary care physician	Primary care, preventive health, management of medical complications in late stages
Dentist	Aggressive, proactive dental care
Hospice care team	Comfort- and dignity-directed care in terminal stages

Current Therapeutic Options for Huntington's Disease: Good Clinical Practice Versus Evidence-Based Approaches?

Killoran, A, Biglan, K *Movement Disorders*, Vol. 29, No. 11, 2014



Middle Stage



- Individuals might experience an increase in their movements and will likely require medication to attenuate movements.
- Complex tasks more challenging :
 - Planning and organization may be difficult
 - Shortened attention span
 - Slowed processing speed
 - Less motivated
- Driving might become an area of concern and debate.
 - **Tough issue** – can be the “elephant in the room”
 - Reevaluate on a regular basis
 - Remain open and honest with your physician
 - Might require a formal or informal driving assessment
 - Safety is of utmost importance!

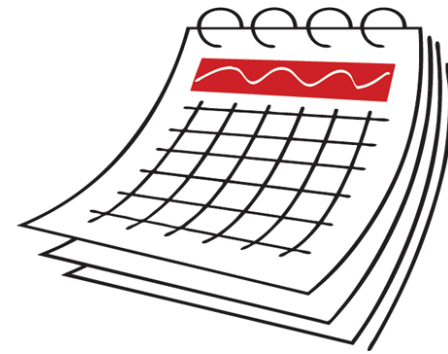
Driving and HD

- Driving is complicated...
- Rebock et al. assessed neurological and cognitive impairments in relation to automobile driving.
 - 73 HD patients (pts)
 - 53 (72%) continued to drive after onset of illness.
 - 29 HD pts still driving and 16 controls were assessed.
- HD pts performed significantly worse on the driving simulator tasks and were more likely to have been involved in a collision in the preceding 2 years. (58% of HD vs 11% of controls).
- Pts who had collisions were less functionally impaired, but had slower simple reaction time scores.
- Summary – HD pts are at increased risk for accidents, but pts who have accidents are not easily distinguished from those who do not.
- Ross et al. Determinants of Functional Disability in HD: Role of Cognitive and Motor Dysfunction. Movement Disorders, Vol.29, No. 11 2014

Middle Stage

- Medications may be necessary to address movements.
 - Ex.- Tetrabenazine or neuroleptics for chorea
- Medications available for behavior or mood disturbances.
 - Ex.-escitalopram, paroxetine, sertraline
- Lifestyle modifications may be necessary.
- Change in level of involvement with occupation..
- Physical therapy
- Occupational therapy
- Speech therapy
- Remain active - continue to exercise!

Tips for caregivers...



- Planning and organizational challenges
 - Make lists that help organize necessary tasks
 - Provide the person with HD with consistent, predictable schedules and routines
 - Maintain a highly structured environment each day
 - Offer limited choices and avoid open ended questions
 - Use shorter sentences with 1-2 pieces of information
 - Establish a central location for posting a daily schedule
 - Consider a daily planner

Tips for caregivers...

- Difficulty with attention
- Try to decrease distractions by turning off TV, radios and phones while completing a task
- Help the person with HD do only one task at a time – true for all of us



Late Stage (TFC 0-2)

- Characterized by the need for 24 hour supervision and care.
- Individuals are dependent on others for their ADLs (personal hygiene, feeding, mobility, dressing).
- Communication may become more challenging.
- Swallowing may become more difficult.
- Motor features may change.
- May last up to a decade.
- Multidisciplinary team is imperative.

Nursing Home Placement

- Rosenblatt et al. examined 799 HD patients at the Baltimore Huntington's Disease Center for features that contributed to nursing home placement.
- 88 of the pts were relocated to care facilities during the 9.2 years of follow up.
- Psychiatric symptoms did not significantly predict institutionalization.
- Level of independence with ADLs was a strong predictor.
- Neither age at onset nor CAG repeat length number alone were significantly correlated with disease duration at the time of institutionalization.
- Studies did not include caregiver stress.

Late Stage HD and Motor Features

- Chorea – may persist throughout the course
 - Often peaks roughly 10 years after onset
 - As chorea declines, dystonia increases
- Dystonia –
 - Involuntary sustained contraction of agonist/antagonist muscles
 - Described as twisting, turning, or cramping
 - Can cause pain
- Parkinsonism
 - Rigidity - stiffness
 - Bradykinesia – slowness of movement
 - Postural instability – difficulty protecting your posture/balance

Late Stage HD and Treatments

- Chorea – Non medical treatments
 - Lower the bed (mattress on the floor)
 - Avoid restraints
 - Weighted utensils
 - Padded bedrails
 - Wheelchairs/Padded seats – Broda or Q foam chair
 - Avoid long cords
 - Frequent skin assessments

Late Stage HD and Treatment

- Gait disturbance and frequent falls –
 - Protective gear
 - Assistive device (walker with caution)
 - Wheelchair earlier rather than later – may increase ability to participate in outings.
 - May need a modification in medications (increase or decrease dose).
 - There is not a medication that directly addresses gait issues (although indirectly medications may help).
- Physical therapy for transfer and range of motion.
- SAFETY FIRST!

Late Stage HD and Treatment

- Chorea –
 - Tetrabenazine
 - Neuroleptics
 - Amantadine
- Dystonia –
 - Oral medications
 - Botulinum toxin injections
- Parkinsonism –
 - May need to try carbidopa/levodopa or dopamine agonists in low doses

Late Stage HD and Treatment



- Oral motor dysfunction results in -
- Dysphagia (difficulty swallowing) – risk for aspiration pneumonia
 - Speech therapy
 - Chin tuck
 - Thickeners
 - Straws
 - ?Feeding tube
- Drooling – botulinum toxin injections can help
- Dysarthria – speech therapy
 - Reduced communication -> mutism – communication board
- Weight loss – frequent high calorie snacks



Late Stage HD

- Cognitive decline
- How does HD differ from Alzheimer's disease (AD)?

Ability	Huntington's Disease	Alzheimer's Disease
Speed of processing	Slow, but relatively accurate	Slow, often inaccurate
Speech output	Slurred and slow, but accurate	Normal in clarity and rate; often the incorrect word
Learning new information	Disorganized and slow, but can learn	Rapid forgetting, defective storage of information
Free recall of memory	Impaired, cannot find the right word; can recognize with choices, benefits from cues	Impaired, memory store is defective; cannot recognize, cues don't help
Motor memory	Impaired, cannot learn or recall motor memories	Intact, can learn and retain motor memories

Cognition and HD

- No approved medications for cognition decline in HD YET
- Important to have uncomfortable conversations early
- Discuss advance directives
- Assign a medical power of attorney

Tips for Caregivers...

- Allow additional time for the person with HD to respond to questions
- Try yes or no questions to speed a response from a person with HD
- Use short, simple sentences and assess understanding frequently
- Remember that even if the person with HD is unable to speak, they can understand what is being said

Tips for caregivers...

- Spatial perception challenges
 - Pad furniture with sharp corners
 - Carpet the floor
 - Keep clear pathways if/when possible

Behavioral Symptoms and Late HD

- Depression
- Anxiety
- Impulsivity
- Perseveration
- Obsessive compulsive symptoms
- Paranoid delusions
- Hallucinations

Strategies for coping...



- Impulsivity -
- It is important to let the person with HD know that yelling is not the best way to get attention and offer alternative methods
- Remember - It is often the “HD talking”, try not to take it personally
- A predictable daily schedule can reduce confusion and stress
- Use deep breathing techniques

Strategies for coping...

- Irritability
- Caregivers should learn the signals (verbal or nonverbal) that show the individual is upset
- Attempts should be made to avoid situations that trigger frustration
- The environment should be kept as calm as possible
- Communicate concerns with the MD

Late Stage HD and Treatment

- Depression/anxiety
 - Effective medications
 - ex. SSRIs, TCAs
 - Psychotherapy
- Obsessive compulsive behaviors and impulsivity
 - Behavioral modification
 - Identify triggers
 - Redirection
 - Medications
 - Antipsychotics
 - Mood stabilizing agents – ex. valproic acid

Late Stage HD

- Possible complications of advanced HD:
- An acute change in symptoms is likely not HD, but a complication of HD.
- Such as:
 - Aspiration pneumonia
 - Urinary tract infection
 - Subdural hematoma
- Deep vein thrombosis - > pulmonary embolus
 - Prevention measures
 - Watch for leg swelling or pain

Late Stage HD and Treatment

- Communication –
 - Keep lists for caregivers of patients likes/dislikes
- Strategies for caregivers...
- Pts might have difficulty with time perception
 - Use frequent and gentle reminders to keep on schedule
 - Avoid time pressure if possible
 - Allow extra time if possible
- Apathy/withdrawal
 - Medications may help
 - Can be due to depression and will respond to antidepressants
 - If not related to depression, stimulants might help.

Late Stage HD

- Maintain an appropriate sleep wake cycle
- Circadian rhythm disturbances can contribute to confusion
- Minimize daytime napping
- Encourage activities during the day
- Practice sleep hygiene
- Avoid sedating medications during the day

Advanced HD Functional Capacity Scale					
	Mobility	Feeding	Continence	Communication	Participation
4	Walks, may have missteps, but no more than occasional falls			Communicates with people other than family, caregivers	Able to participate actively in family/residence activities
3	Frequent falls or very frequent near falls	Eats independently, using at least a fork or spoon	Continent of both stool and urine, takes care of toilet hygiene	Phrases or sentences only intelligible to family or in context	Able to participate in some or occasional activities
2	Wheelchair, independent	Uses a cup/straw, finger foods	Not always continent, or poor toilet hygiene	Single words or severe dysarthria; limited ability to speak even with family or in context	Able to attend some activities but little or no active participation
1	Able to sit but dependent on others for mobility	Must be fed	Incontinent most or all of the time but aware	Mute but attempts to communicate (grunts, screams)	Able to respond interactively in some way to others
0	Bed-bound or unable to sit	Most or all nutrition provided by feeding tube	Incontinent and unaware or passive about help	Mute	Non-interactive

Maximum score 18 points, minimum score 0 points

Stage A1: 16-18 points (largely independent)

Stage A2: 10-15 points (still trying to be independent but not very successful)

Stage A3: 6-9 points (receives assistance with all activities but at least interacts)

Stage A4: 2-5 points (requires full assistance with everything)

Stage A5: 0-1 points (terminal stages, mute, bedbound, non-interactive)

Tips for Caregivers...

- During an outburst, caregivers might need to:
- Redirect the HD person away from the source of anger
- Acknowledge the individual's irritability and frustration
- Try not to take the insults personally
- Leave the room if the threats become violent

Terminal Stages of HD

- Patients may begin to scream .
 - Evaluate for any treatable sources
- Excessive sleeping might also be a clue.
- Hospice/palliative care
 - Symptomatic treatments can continue.
 - **The focus is on comfort/quality of life for the patient**
 - This does not mean you are giving up.
 - Can provide respite for caregivers.
 - Maintain our patients' dignity while dying.

Summary

- HD is an inherited progressive neurodegenerative disease.
- There are multiple available therapies (medical and nonmedical).
- It takes a village – multidisciplinary teams are important.
- The goals for treatment should be maximizing functional abilities and optimizing quality of life.
- Fantastic research is currently underway.
- Thank you for your time.

- **H**ope is important – TEAM HOPE!
- **U**nderstand the disease – knowledge is power!
- **N**urture your strengths.
- **T**ake your time.
- **I**nvoke others to learn about HD – raise awareness!
- **N**ecessitate continued advancements in treatment - research!
- **G**et up and move – exercise is therapeutic!
- **T**reat yourself once in a while (caregivers included)!
- **O**pen up to others and ask for help!
- **N**ever give up!!
- **S**tand up for HD, continue the fight!

Resources

- www.clinicaltrials.gov
- www.wemove.org
- www.HDSA.org
- <http://en.hdbuzz.net/>
- www.huntington-study-group.org