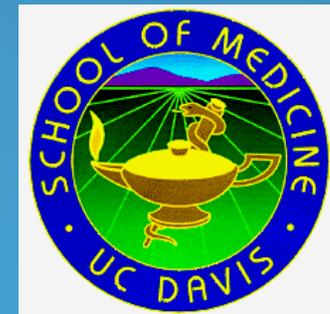


Managing early/mid-stage HD: Behavioral and medication strategies

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HDSA Center of Excellence at UC Davis

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Presenter Disclosures

Dr. Vicki Wheelock

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

No relationships to disclose
or list



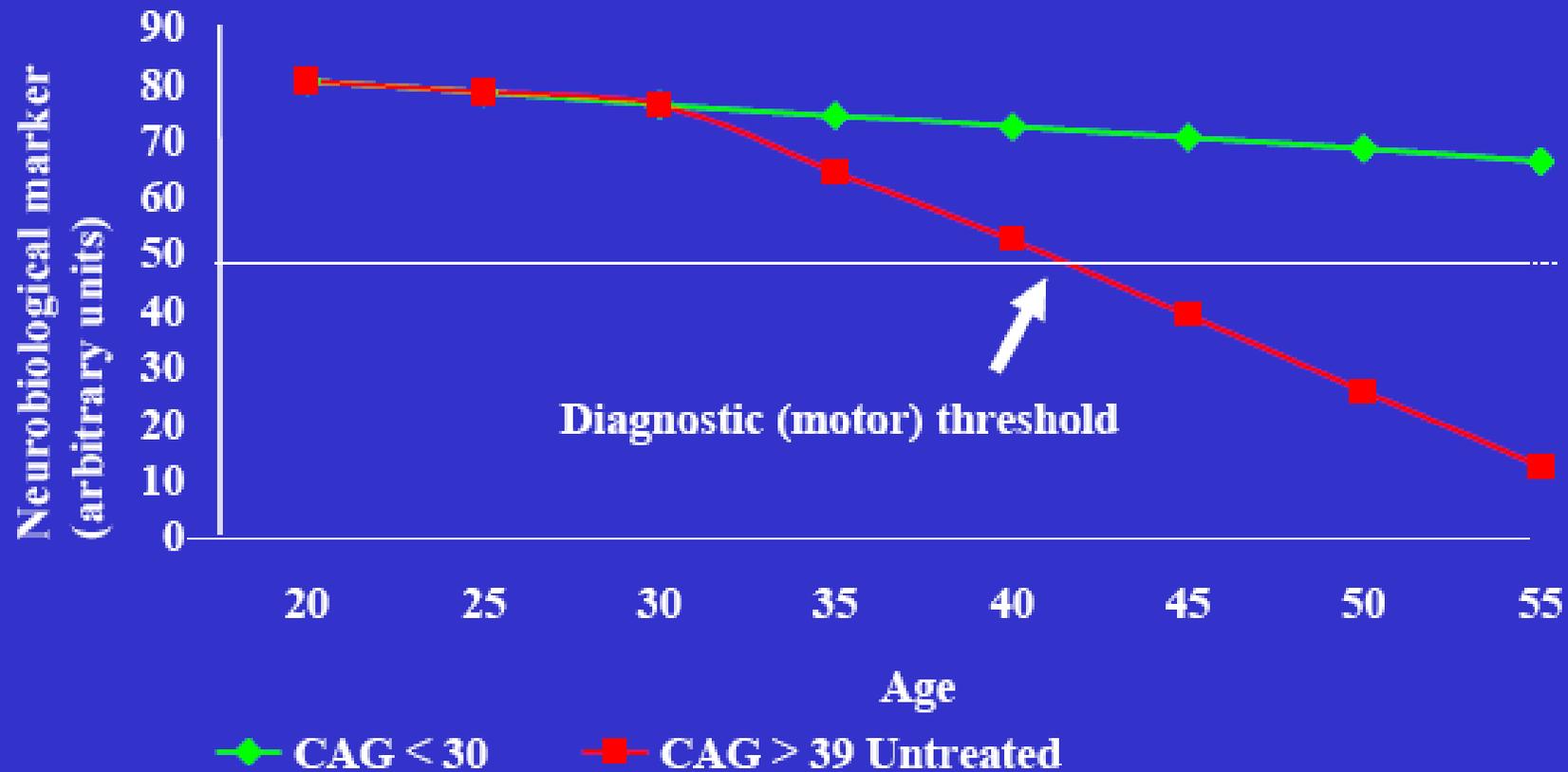
Huntington's Disease Society of America



Outline

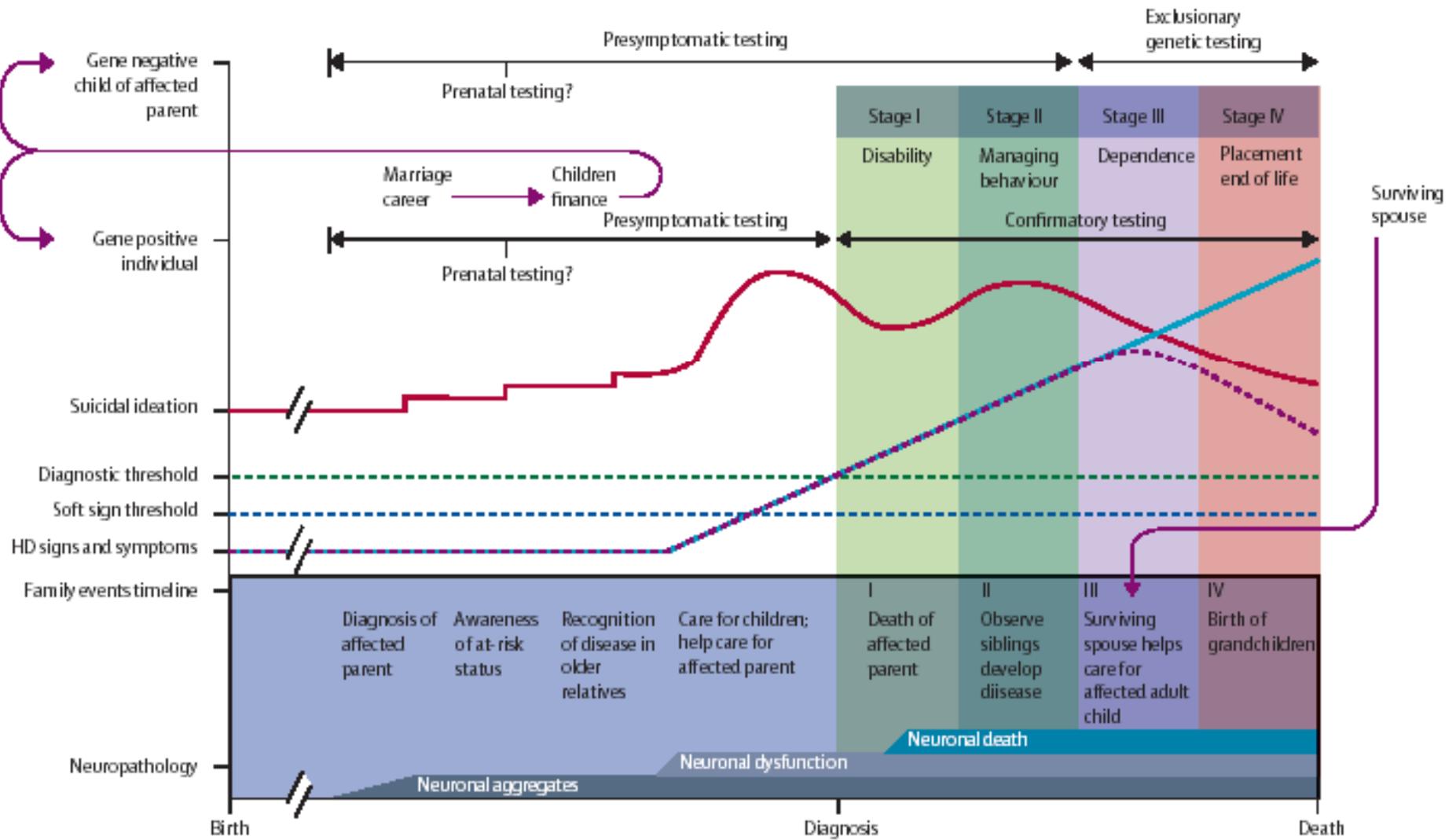
- Definition of HD stages
- HD triad: motor, cognitive and emotional
- Trajectory: from stage to stage
- Scenarios to consider
- Resources

HD Over the Lifetime



Paulsen JS, Hayden M, Stout JC and the PREDICT-HD Investigators.
Preparing for clinical trials: The PREDICT-HD study. *Arch Neurol* 2006;63:883-890

Life cycle in HD





Huntington Disease Stages

Stage 0: Presymptomatic

Stage 1: Slightly lower performance at work; independent at home

Stage 2: Can still work (lower level), still mostly independent at home

Stage 3: Difficult to work, starts to need help with financial, home activities

Stage 4: Unable to work. Needs major assistance with care

Stage 5: Full-time nursing care required

Adapted from Shoulson et al, Quantification of Neurological Deficit, Boston:Butterworth,1989

Total Functional Capacity (TFC) Score

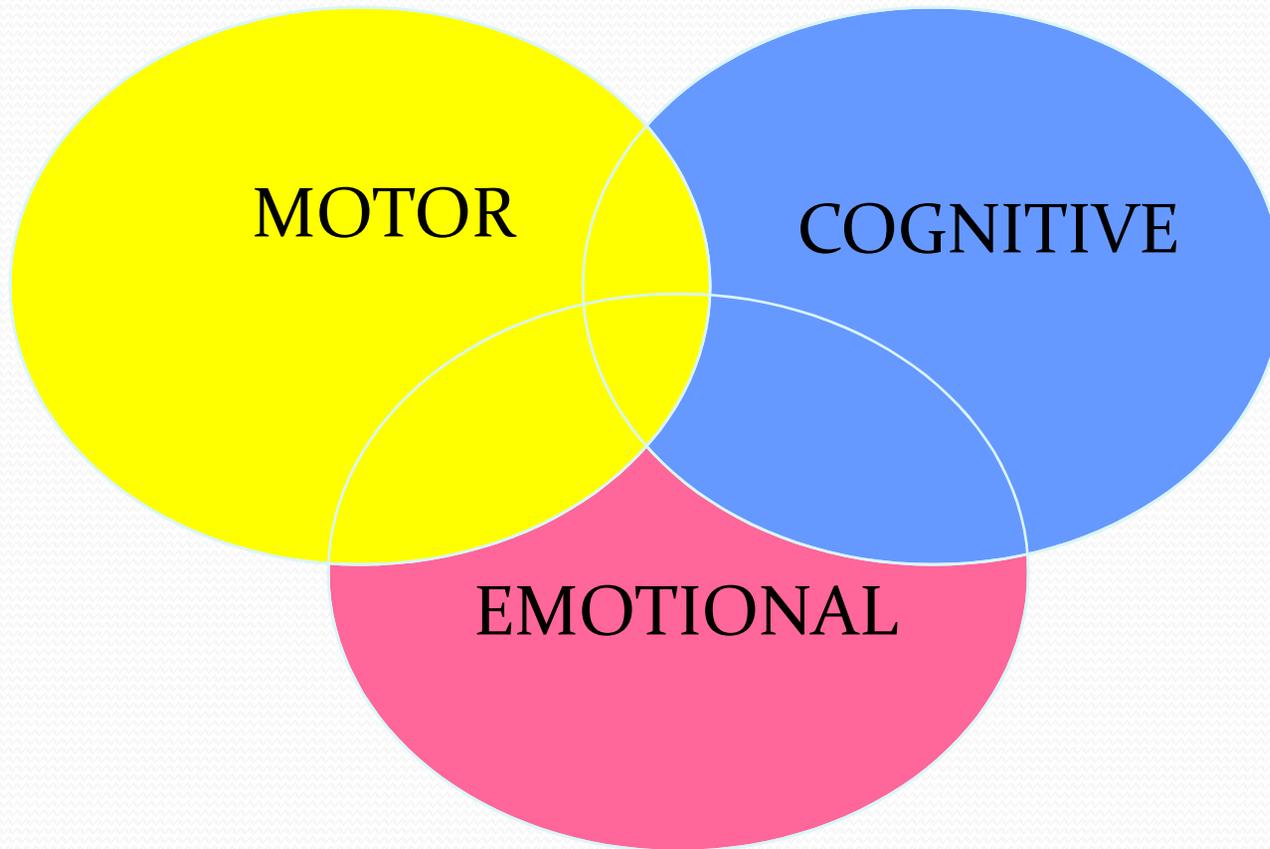
Abilities to:	Points
Work	0 - 3
Handle finances	0 - 2
Do home activities	0 - 3
Perform self-care	0 - 3
Live at home	0 - 2
TOTAL Score	0 - 13

HD Staging	Points
Stage 1	11 - 13
Stage 2	7 - 10
Stage 3	4 - 6
Stage 4	1 - 3
Stage 5	0

TFC declines by 0.7 units/year

Marder et al, Neurology 2000;54;452-8

Symptoms in HD



Meanwhile, in the brain.....

Behavior

Multi-tasking

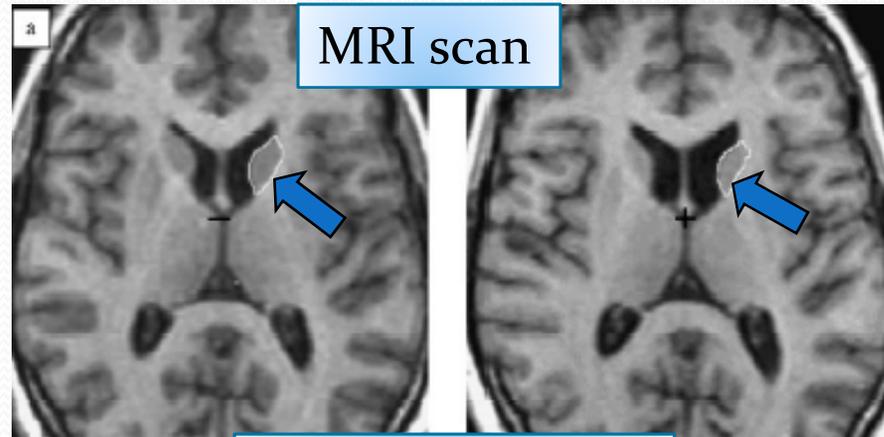
Episodic anger
Irritability

Impulsivity

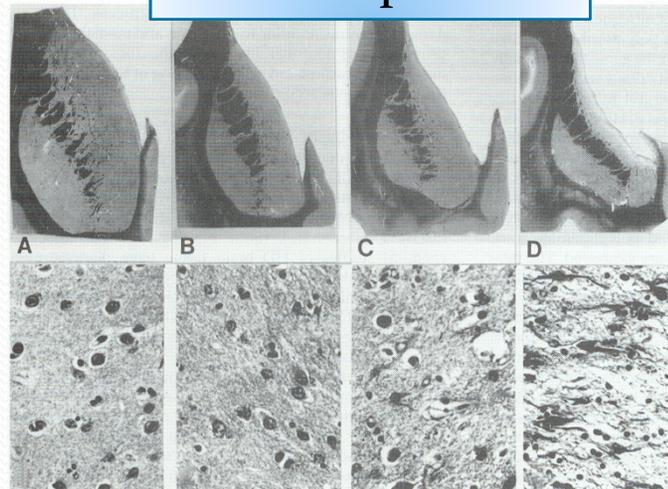
Changes in
sleep/wake cycle

Organizing
Concentrating
Prioritizing

Aylward EH. Brain Research Bulletin 2003;62:137-141



Microscopic view



Stages 1 2 3 4

Movement

Chorea:
involuntary
movements

Restless, fidgets

Slowness of
movement

Balance problems

Fine motor tasks

Emotional and psychiatric problems in HD

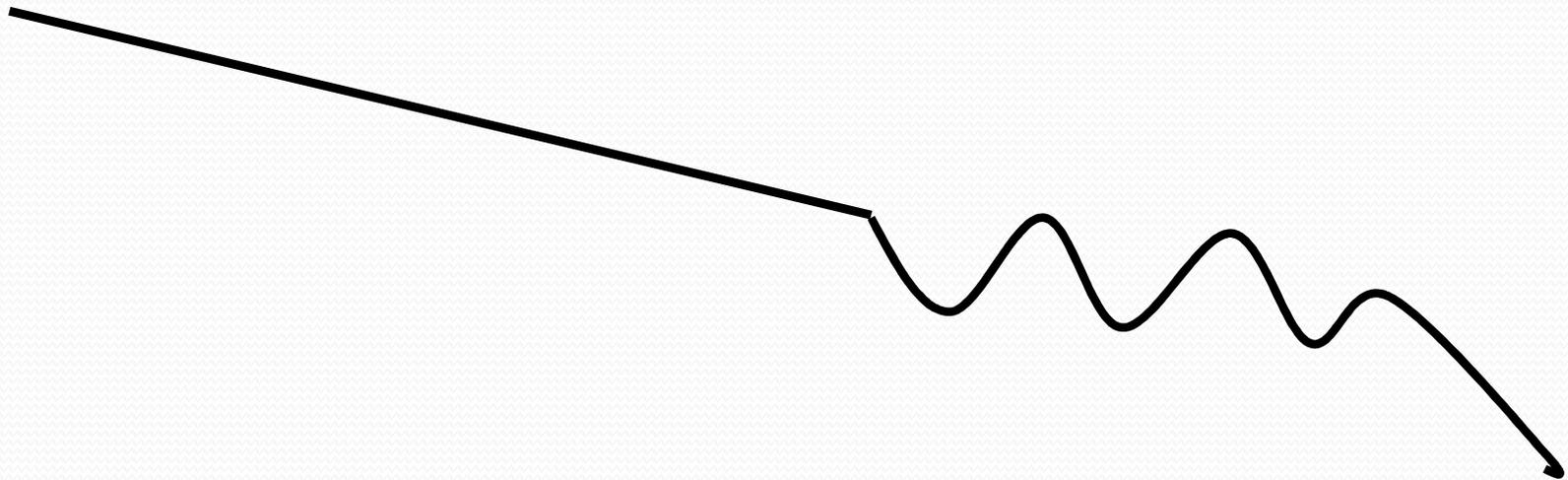
Symptom	Prevalence
Depression	40 – 80%
Anxiety	30 – 40%
Obsessive/compulsive disorder	10 – 20%
Irritability	common
Apathy	20%
Episodic anger	common
Psychosis	5%



Treatment involves much more than medication...

- Autonomy
- Dignity
- Meaningful social interaction
- Communication
- Comfort
- Safety and order
- Spirituality
- Enjoyment, entertainment and well-being
- Nutrition

Managing the Trajectory



Source: T. Tempkin RNC, MSN, ANP



Scenario 1

A 38 year old woman who tested positive for the HD CAG expansion 5 years ago comes to the clinic to establish care. She has never experienced chorea or clumsiness, but she has had persistent feelings of sadness and hopelessness, sleeps poorly, and has lost weight. She moved from one job to another in the 2 years, and gives vague reasons for the job changes. She admits that she's missed paying some bills on time, and that she doesn't keep her apartment as clean as she used to. She wonders if she might be starting to get symptoms of HD.



Managing the Trajectory: Stage I

- Still working– but may be stressed or starting to decline in performance
- Still independent at home, but multi-tasking begins to become more difficult
- Emotional symptoms of anxiety, depression, irritability may start

- Stress management
- Changes at work
- Allow more time; avoid multi-tasking
- Seek treatment for cognitive and mood changes



Medications for depression and/or anxiety

- Cognitive-behavioral therapy can be very effective
- SSRI drugs (Zoloft, Paxil, Celexa for instance)
- NSRI drugs: Welbutrin, Effexor
- SNRI: Cymbalta
- Start with low doses, may be continued for several months. Dose can be lowered and drug discontinued for some patients; others do well with long-term treatment



Scenario 2

A 28 year old man with the HD CAG expansion gets angry at work and assaults a co-worker. He quits that job and is re-hired at another, and no legal problems have surfaced. He denies difficulty with irritability, but his wife reports frequent anger outbursts at home. His neurological exam shows only questionable signs of HD.



Managing irritability and anger

- Establish and stick to routines, schedules
- Look for behavioral triggers; re-direct
- Avoid alcohol, stimulants like caffeine
- Avoid confrontations and ultimatums
- Consider anxiety or depression as a cause
- If anger is severe and/or frequent, medications will help
- Call authorities if necessary

Medications for irritability and aggression

- First line: SSRIs
- Second line: mood stabilizers: anti-seizure medications like Trileptal, Depakote, others
- Third line: mood stabilizer anti-psychotic meds like Zyprexa
- Obsessive thoughts and compulsive behaviors
 - SSRI anti-depressant medications help



Scenario 3

A 25 year old woman is brought to the HD clinic by her father. Her mother died of HD in her 30's, and her older sister was diagnosed with HD a few years ago. She has had difficulty keeping a job for the last year or so, has become more withdrawn and apathetic. Her neurological exam shows that she has mild memory problems, marked slowness in movement, rigidity in her arms, poor balance.



Managing the Trajectory: Stage II

- Can still work – but at a reduced capacity.
- Still mostly independent at home, but organizing and prioritizing may be more challenging, and may not be as engaged in family activities.

- If possible, negotiate with work place for modified duties
- Begin maintaining a schedule
- Seek treatment for cognitive and mood changes
- Future planning
 - Disability
 - Long-term care

Medications for Juvenile HD or adult rigid/dystonic HD

- Muscle stiffness, rigidity, slowness, tremor, loss of coordination, not typically chorea
- Treatments:
 - Baclofen
 - Diazepam
 - Parkinson meds: levodopa, amantadine
 - Botox injections for local therapy
- Physical therapy
- Occupational therapy
- Adaptive equipment



Medications for behavior changes

- Apathy
 - Must first distinguish apathy from depression
 - Always make environmental changes: provide structure, schedules, reminders.
 - If above fails, consider using stimulant drugs such as Ritalin (methylphenidate)
- Cognitive difficulties
 - More likely to be a problem in people with younger age at onset
 - Can consider using drugs developed for Alzheimer's disease (donepezil, memantine)



Scenario 4

A 43 year old ice-cream truck driver whose father died of HD is the sole support for his family. He immigrated to the US from another country 4 years previously. He's had no behavioral problems, but now needs help handling money. His examination shows severe chorea. His wife doesn't drive, and he won't qualify for Social Security because he hasn't worked long enough in the US.

Managing the Trajectory: Stage III

- Difficult to work/may stop working.
- Needs help with finances and home activities.

- Financial planning and assistance
- Routine, routine, routine..... 😊
- Driving cessation
- Maintain social contacts and stimulation.
- Balance the burden with.....Life!



Medications for chorea

- First-line: Tetrabenazine (FDA-approved in 2008 as the first drug in US for HD)
- Second line: benzodiazepines
- Third line: antipsychotic drugs
 - “Typical” (older): haloperidol, fluphenazine
 - “Atypical” (newer): olanzapine, risperidone, others

Tetrabenazine



Benefits: reduces chorea

Side effects:

swallow dysfunction
depression/suicide
restlessness

Interactions:

Some anti-depressants
Other anti-chorea drugs

Dosing:

Genetic test
Monitor EKG

Cost: Special program



Scenario 5

A 59 year old psychologist comes to clinic about 5 years after she developed chorea, and 2 years after she retired from her practice. Her father died of HD; she thinks she has it, but has avoided seeking medical care because “there’s nothing you can do anyway.”

She has frequent falls, severe chorea and poor balance.

Genetic testing confirms the diagnosis of HD. Her chorea improves with medications, but she grows more irritable, impulsive, and begins to lose weight. She often refuses to let her caregiver help her with personal care, and her hygiene declines.



Managing the Trajectory: Stage IV

- Unable to work.
- Needs major assistance with personal care

- Ongoing needs for financial assistance/planning
- Medical treatments can address problems with swallowing, movement, gait, balance, weight, mood and cognition.
- Family caregivers need more support.



Management of weight loss

- Nutrition/dietary consult
- Swallow eval by speech therapy
- Increase calorie intake
- Treat chorea
 - Tetrabenazine is FDA approved
 - Zyprexa is also effective, and causes weight gain as a side effect)
- Maintaining ideal body weight (or slightly above) may help mood, behavior, motor control



Scenario 6

A 29 year old successful young professional developed psychosis as the initial manifestation of HD. Her psychosis is well controlled with medications. By age 36 she can no longer work, but externalizes the reason. Her husband divorces her, she no longer has a car, and she comes to clinic expressly to “get my car keys back.” She insists that she doesn’t have HD.



Unawareness and HD

- Not the same as psychological denial
- It's quite common, but not everyone with HD has this difficulty
- Can be limited to some symptoms and not others
- Can lead to work, family conflict, injury
- Treatment is difficult; behavioral strategies are best

Patients with Huntington's disease have impaired awareness of cognitive, emotional, and functional abilities. Hoth, Paulsen et al, J Clin Exper Neuropsychol 2007: 365-376.

Driving and HD

Single study: Johns Hopkins, 1995

- *(Rebok et al, Mov Disord 1995;10;778-87)*
- 73 HD patients, 52 still driving
- 29 HD patients and 16 Non-HD controls underwent driving simulator evals
- Findings:
 - HD accident rate 58% over 2 years
 - Controls: 11% over 2 years
 - HD patients had slower reaction times, but HD stage did not correlate with risk of accidents



Driving and HD

- Experience, judgment, attention, reaction time, visual-motor integration and motor skills all contribute to driving ability
- Medications (esp. antichorea) can cause sedation
- No single predictor of accidents exists
- Reporting laws vary by state
- Do ask patient and family about driving difficulties
- Do recommend HDSA ID card and Medic-Alert bracelet for those still driving
- Recommend driving eval if driving skills in question



Scenario 7

A 45 year old man is brought to the HD clinic by his family. He was diagnosed with HD 2 years ago, and has become much more difficult at home. He has frequent anger outbursts, flies into rages, believes that his neighbors are planning to rob him, and is restless and can't sleep. His family is afraid that he may hurt himself or others.



Managing psychosis

- Seek the help of a psychiatrist
- Ensure safety of the patient and others
- Anti-psychotic meds will help
 - Typical agents
 - Atypical agents
- May need hospitalization to assess and start treatment.
- Call the authorities if patient refuses evaluation or treatment



A few final words

- It's never too early to start behavioral strategies
 - Structure and routines help tremendously
 - Use calendars
 - The HD mantra: one thing at a time
 - Regular schedule of sleep/wake, meals, activities
- Good for the brain:
 - Healthy diet
 - Regular exercise
 - Avoid brain toxins.....alcohol, smoking
- Caregiving begins at Stage 1.
 - Ask for help.
 - “Share the Care!”



Thank you to our COE staff and Northern CA Chapter of HDSA!

