



# MANAGING LATE STAGE HD

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# PRESENTER DISCLOSURES

Donald S Higgins, Jr, MD

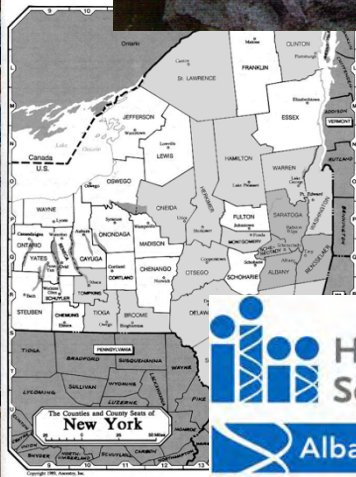
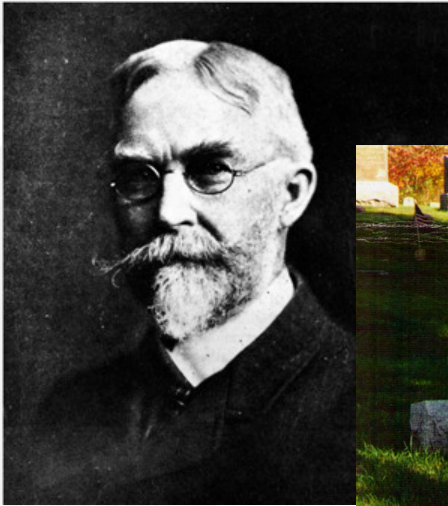
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# UPSTATE NY & HUNTINGTON'S DISEASE



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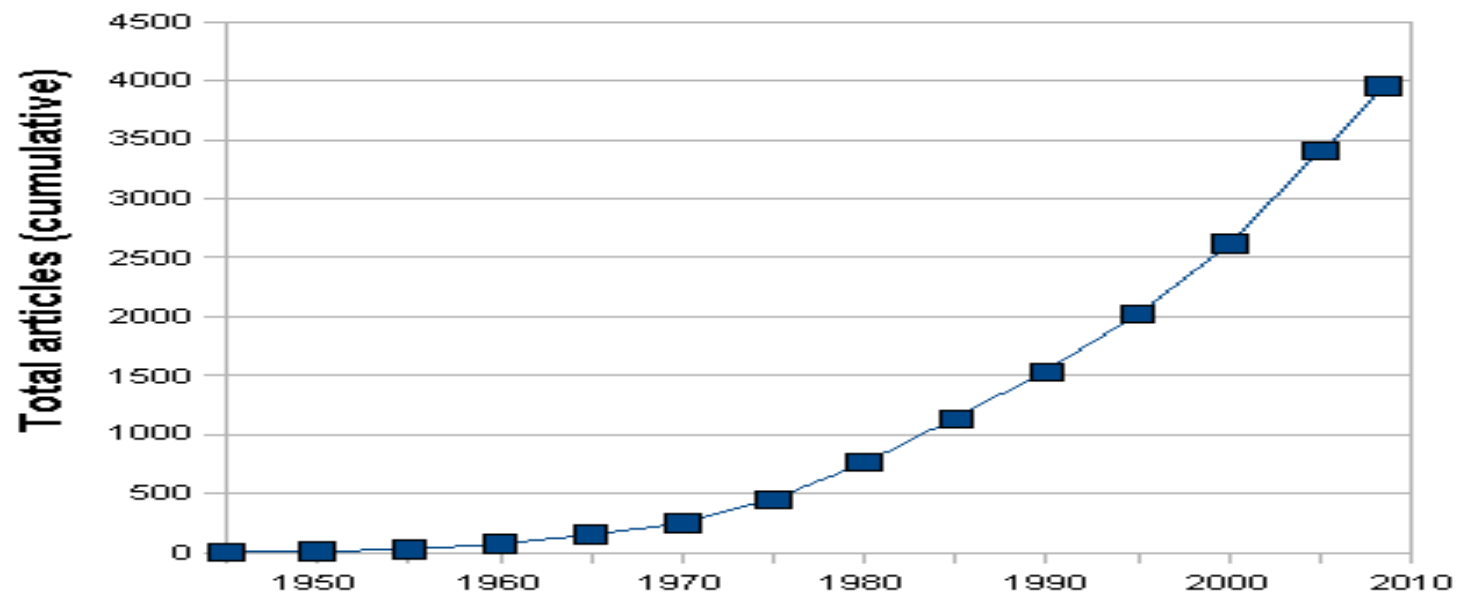
Albany NY/Western MA AFFILIATE



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# RESEARCH

## HD Articles on Pubmed



# TREATMENTS FOR HD

- Palliative

*Reduce the signs and symptoms*

- Preventive

*Slow the onset or progression*

- Curative

*Halt the disease process*





# HUNTINGTON'S DISEASE

- Involuntary Movement
- Psychiatric Disturbance
- Cognitive Decline

# STAGES OF HUNTINGTON DISEASE

**Clinical stages of Huntington's disease (HD)**

Early	Middle	Late
Clumsiness	Chorea (mild)	Chorea (moderate-severe)
Apathy	Dystonia	Parkinsonism
Anxiety	Balance and gait difficulties	Dysphagia
Agitation	In-coordination	Dysarthria
Irritability	Weight loss	Self-neglect
Personality changes	Disinhibition	Dementia
--	Cognitive impairment	Hallucinations/delusions

Ref: Modified from <http://www.ncbi.nlm.nih.gov/books/NBK1305/>



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# STAGES OF HUNTINGTON DISEASE

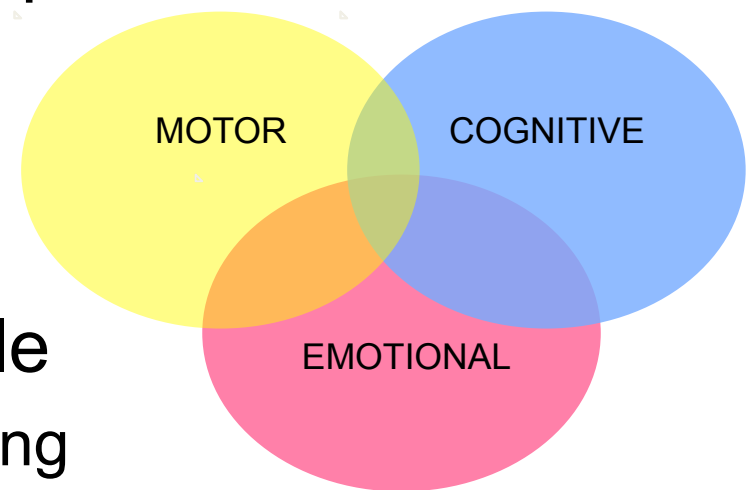
Phase	Years	Symptoms
<b>Transitional</b>	0-3	mood swings behavioral disturbances, hyperreflexia, memory impairment, increased clumsiness, impairment of voluntary movements, eye movement abnormalities
<b>Early</b>	3-5	dysarthria chorea gait abnormalities
<b>Middle</b>	8-10	bradykinesia, rigidity global dementia, dystonia, dysphagia
<b>Late</b>	15-25	incontinence, wasting, aspiration, bed ridden death



	Engagement in occupation	Financial affairs	Domestic responsibility	Activities of daily living	Care provided at...
Stage I TFC 11-13 0-8y	usual level	full	full	full	home
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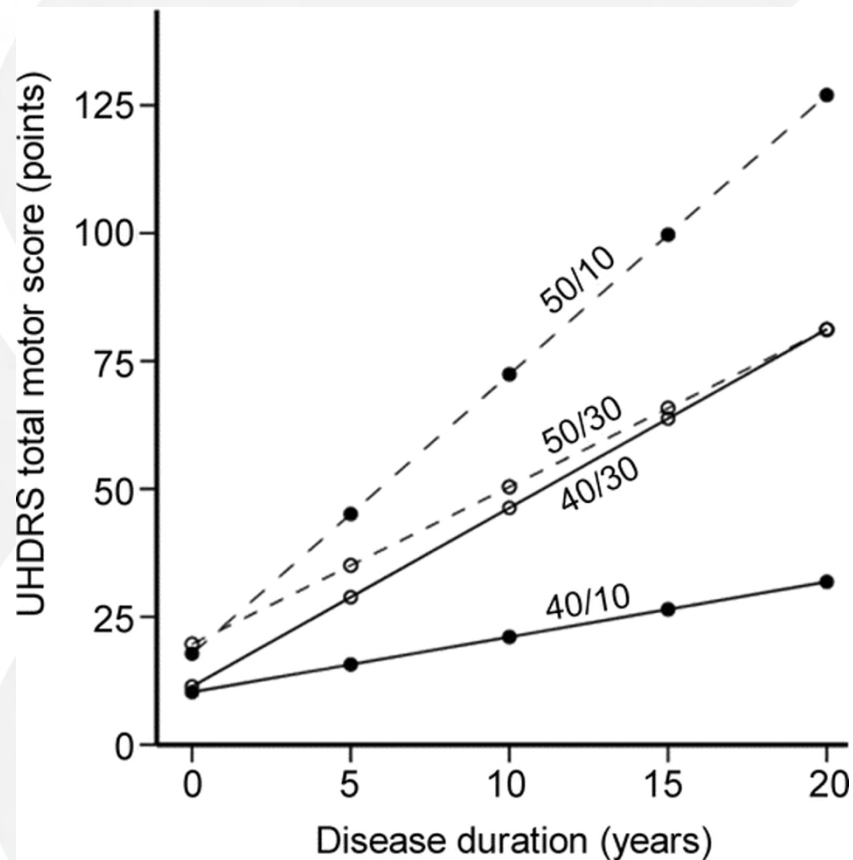
# PRINCIPLES OF MANAGEMENT

- No single treatment for all symptoms
- Target troublesome symptoms
  - Movement
  - Mood
  - Behavior
- Reduce/Stop as possible
  - trial with careful monitoring
  - withdraw if no clear benefit

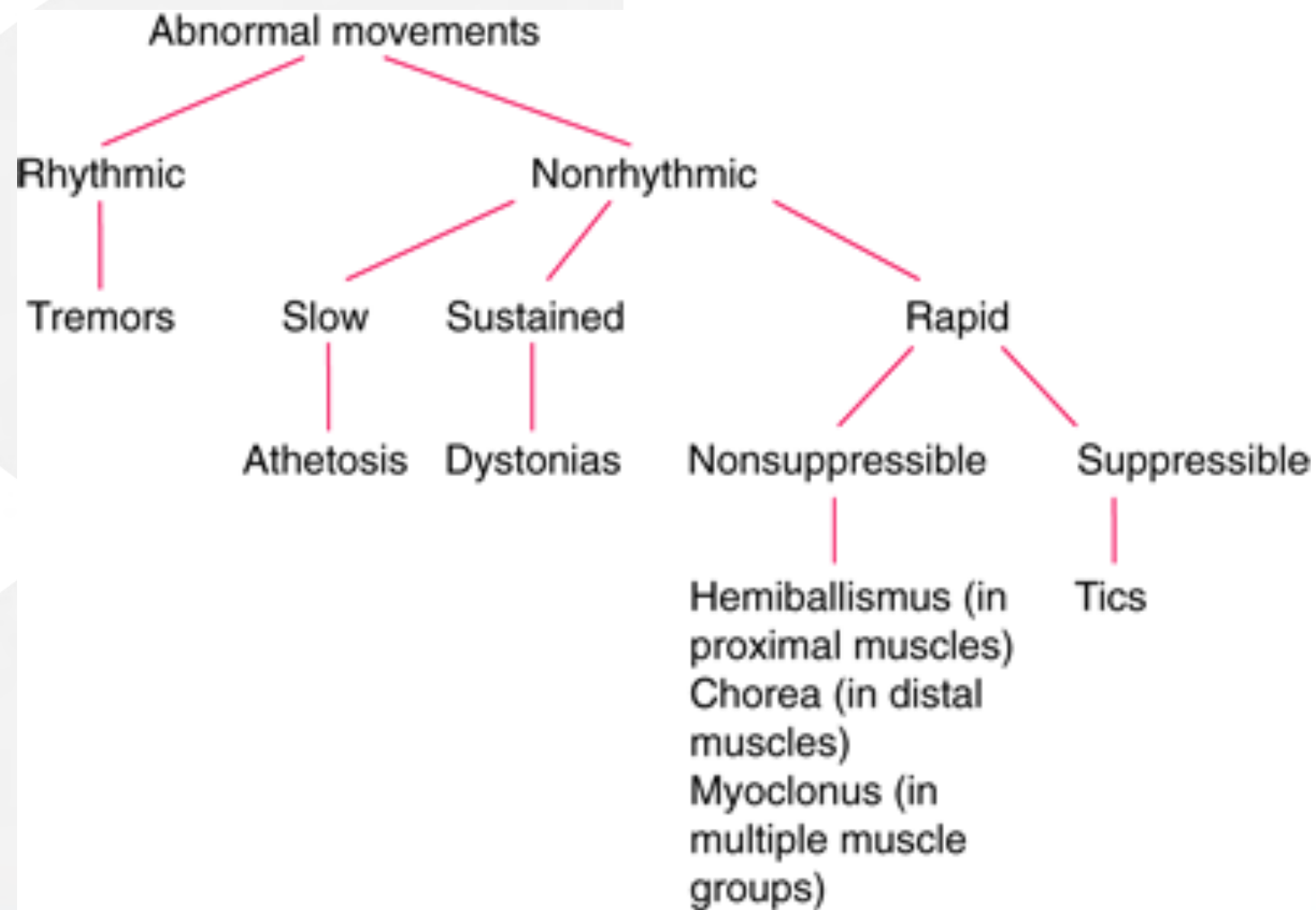


PRIMUM NON NOCERE

# INVOLUNTARY MOVEMENTS



- Neuroleptics/Antipsychotics
  - Typical – Haloperidol (Haldol®)
  - Atypical – Aripiprazole (Abilify®)
- Dopamine depleting agents
  - Tetrabenazine (Xenazine®)
- Benzodiazepines
  - Clonazepam (Klonopin®)
  - Diazepam (Valium®)
- Miscellaneous
  - Amantadine (Symmetrel®)



# INVOLUNTARY MOVEMENTS

- Rigidity/stiffness
  - Advanced disease
    - Anti-parkinsonian medications
  - Amantadine (Symmetrel®)
  - Carbidopa/Levodopa (Sinemet®)
- Dystonia
  - Clonazepam (Klonopin®)
  - Botulinum toxin (Botox®, Myobloc®)



# BALANCE AND FALLS

- Can be early symptom
- Causes:
  - Involuntary Movements
  - Medication
  - Environment
- Management:
  - Slow down!!!
  - Lighting
  - Footwear





# REHABILITATION SERVICES

- Physical Therapy
  - Strength
  - Balance
  - Gait assist device
  - Seating

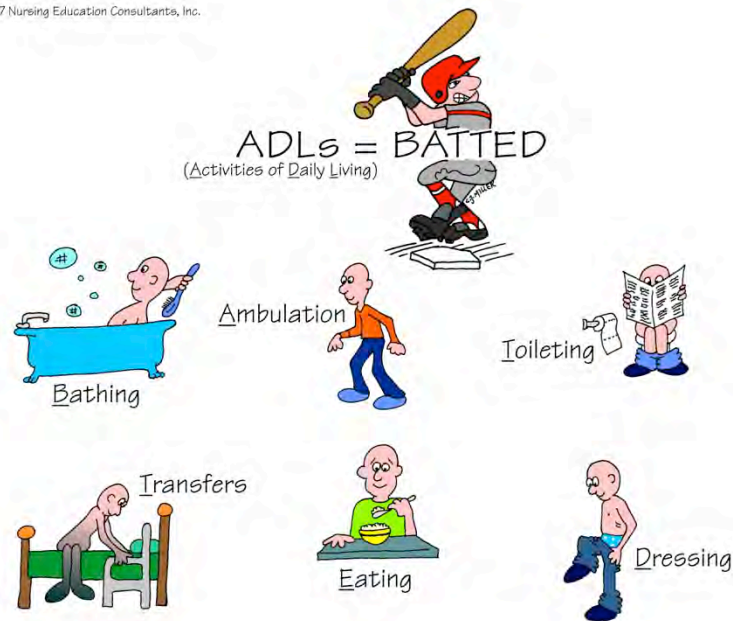


# AMBULATION ASSIST DEVICES



# REHABILITATION SERVICES

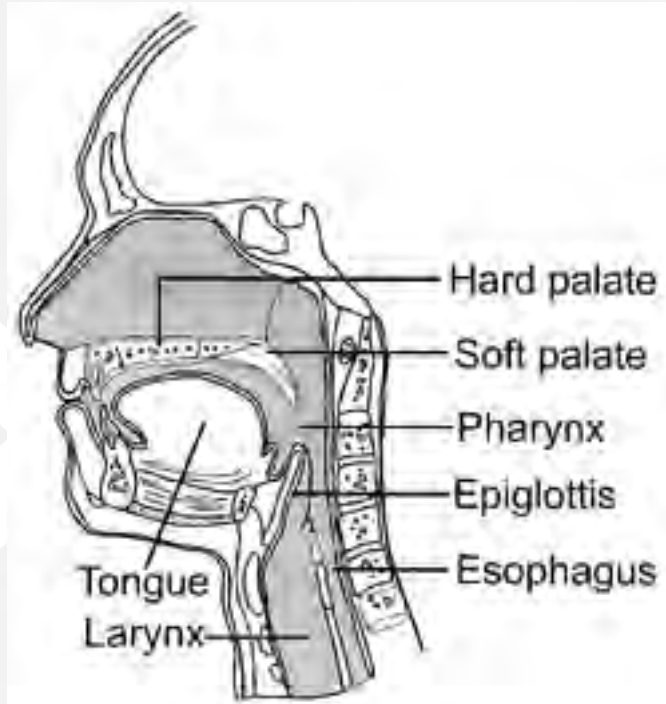
© 2007 Nursing Education Consultants, Inc.



- Occupational Therapy
  - Activities of Daily Living
  - Devices & Gadgets

# REHABILITATION SERVICES

- Speech/Language pathology



SWALLOWING





# DYSPHAGIA

- Positioning
  - Upright posture
  - Limit backward head
- Avoid distractions
- Control rate of intake
- Alternate bites with sips
- Provide small bites and sips.
- Swallows twice for every bite
- Gravies, sauces, & condiments
- No talking



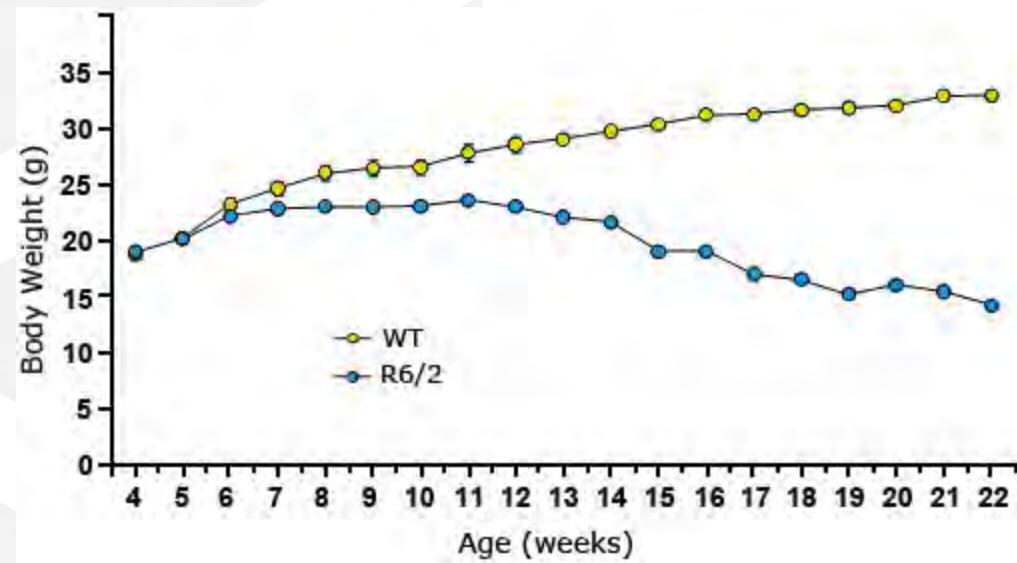
Table 1: Primary cause of death in 147 patients with Huntington disease

Cause of death	N=147	100%
Known Cause:		
-pneumonia	81	55.1
-suffocation	6	4.1
-pulmonary embolism	6	4.1
-cachexia	11	7.5
-cardiac diseases	16	10.9
-other neurological diseases	3	2.0
-shock/sepsis	7	4.8
-suicide	2	1.4
-euthanasia	5	3.4
-other causes	10	6.8

Heemskerk A, Boes RA. PLOS Currents Huntington Disease. 2012  
 † Includes gastrointestinal and urogenital diseases, respiratory diseases

Sorensen SA, J Med Genet 29; 911-914, 1992

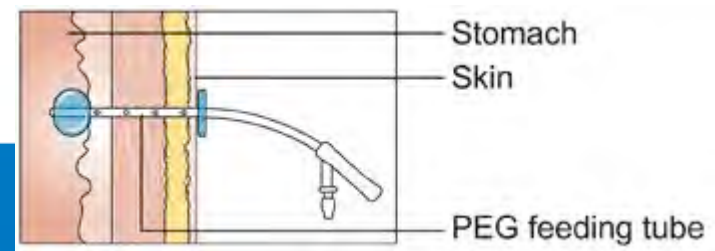
# NUTRITION





# NUTRITION

- Calorie Intake
  - High calorie foods
  - Easy to swallow foods (thickeners)
  - Assistance during meals
  - Multiple meals (5/day)
  - Ready snack access
  - Limit Dairy (increase mucus)
- Medication
  - Medical marijuana
  - Hormonal therapy
- Feeding Tubes
  - PEG
  - J-tube



# COMMUNICATION

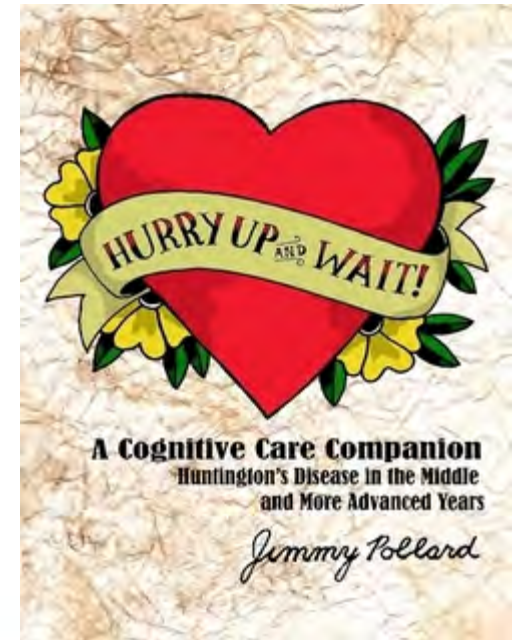
- Eliminate distractions
  - Keep it simple
    - Yes/No
  - One question at a time
  - Gestures/expressions
  - Ask for clarification
    - Repeat what was said
    - Keep to familiar topics
  - Be patient
- Speak slowly
  - Repeat
  - Keep sentences short
  - Speak louder (deep breath)
  - Use alternative techniques
    - Word/alphabet/picture boards
    - Electronic devices

go	you	my	is	where
down	me	to	find	away
said	for	Free Space	help	come
the	we	run	red	a
see	not	little	funny	here



# COGNITIVE IMPAIRMENT

- Timing
- Speed of cognitive processing
- Emotion
- Olfaction
- Memory
- Attentional deficits
- Executive function
- Communication



# COGNITIVE IMPAIRMENT

## Effect of donepezil on motor and cognitive function in Huntington disease

**Abstract**—Striatal cholinergic dysfunction may be important in Huntington disease (HD). We studied whether donepezil improves chorea, cognition, and quality of life (QoL) in HD. Thirty patients were randomly assigned to treatment with donepezil or placebo. At the doses studied, donepezil did not improve chorea, cognition, or QoL. Adverse events were similar between both groups. Based on this small sample study, donepezil was not an effective treatment for HD.

NEUROLOGY 2006;67:1268–1271

E. Cubo, MD, PhD; K.M. Shannon, MD; D. Tracy, MD; J.A. Jaglin, RN; B.A. Bernard, PhD; J. Wu, ScM; and S.E. Leurgans, PhD

- Galantamine (Reminyl®)
- Memantine (Namenda®)

## Rivastigmine in the treatment of Huntington's disease

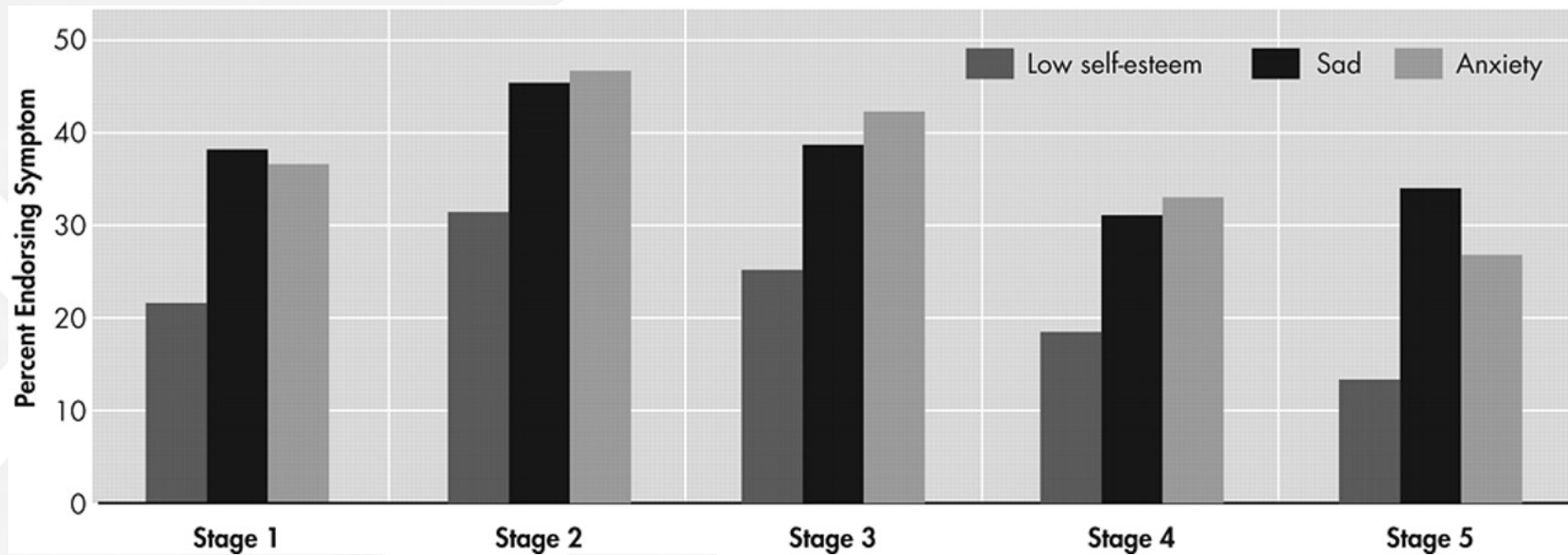
U. Rot<sup>a</sup>, J. Kopal<sup>a</sup>, A. Sever<sup>a</sup>, Z. Pirtošek<sup>b</sup> and A. Mesec<sup>a</sup>

<sup>a</sup>Department of Neurology, <sup>b</sup>Department of Clinical Neurophysiology, Medical Centre, Ljubljana, Slovenia

Correspondence: Uroš Rot, Department of Neurology, Medical Centre, Zaloška 7, 1525 Ljubljana, Slovenia (fax: +386 1431 33 50; e-mail: uros.rot@guest.arnes.si).



# DEPRESSION



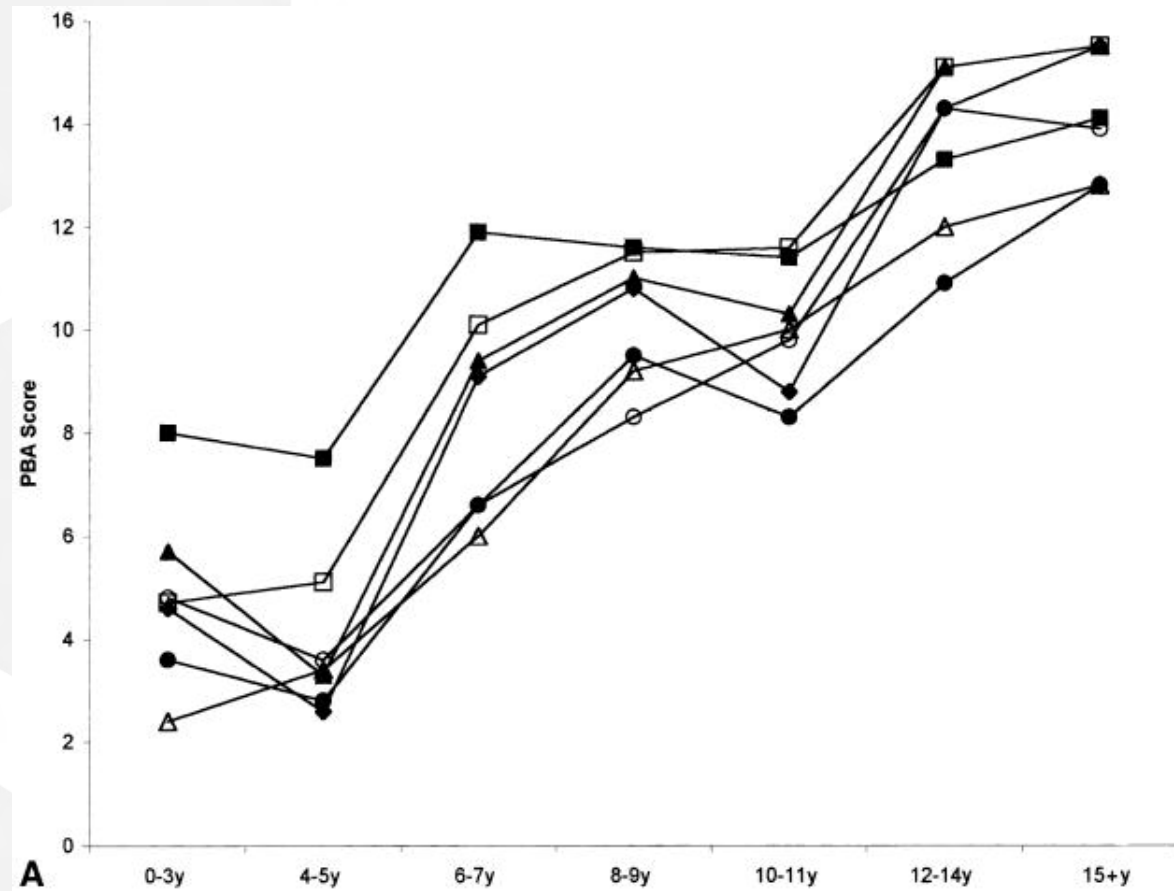
J Neuropsych Clin Neurosci 2005;17:496-502

# BEHAVIOR

## Factor 1: Apathy

Loss of energy  
Failure to complete tasks

Blunting of affect  
Poor self care  
Poor quality of work  
Lack of initiative  
Impaired judgement



Neuropsych Neuropsychol Behav Neurol 2001 Oct-Dec;14(4):219-26

# APATHY

- No established pharmacological treatments
- Education of patient, family & care team
- Psychological measures – structured environment (day care, exercise, etc.)





# AFFAIRS IN ORDER

- Making informed & considered decisions
- Advanced Directives:
  - Living Will
  - Healthcare Proxy
  - Durable Power of Attorney
- Home care
- Assisted care/living
- Institutional care



# END-OF-LIFE DISCUSSIONS

**Table 1** Characteristics of physicians

Respondent (R)/type of physician	Number of patients with HD in the practice	Supportive of euthanasia/participation in euthanasia in HD	Raises the issue of advance directives	Raises the issue of euthanasia with patients
1. GP	1	Yes/yes	Yes	Yes
2. GP	1	Yes/yes	Yes	Yes
3. GP	1	Yes/yes	Reacts to questions	Reacts to questions
4. GP	1	Yes/yes	Yes	Reacts
5. NHP	>100	Yes/yes	Yes	Yes
6. NHP	>50	Yes/yes	Yes	Yes
7. NHP	>50	Yes/yes	Yes	Yes
8. NHP	1	Yes/no	Yes	Reacts
9. NHP	>30	Yes/yes	Yes	Yes
10. NHP	>30	Yes/yes	Yes	Yes
11. NHP	>70	Yes/yes	No, reacts	No, reacts
12. Psychiatrist	>10	Yes/no	Reacts	Reacts
13. Psychiatrist	>50	Yes/yes	No, reacts	Reacts
14. Neurologist	>50	Yes/yes	No, reacts	No, reacts
15. Neurologist	>30	Yes/no	No	No

GP, general practitioner; HD, Huntington's disease; NHP, nursing home physician.

Booij SJ, et al, J Med Ethics 39; 621-624, 2013

# NURSING HOME

**Table 1**  
Demographic Characteristics of SNF Residents With HD

Diagnosis	Total Number	Percent
HD	340	0.14%
Residents in MDS database	249,811	
MDS Data Element	Total Number	Percent
Sex		
Male	134	39.4%
Female	206	60.6%
Age category		
<30 years	1	0.3%
30–34 years	7	2.1%
35–39 years	17	5.0%
40–44 years	15	4.4%
45–49 years	23	6.8%
50–54 years	48	14.1%
55–59 years	51	15.0%
60–64 years	46	13.5%
65–69 years	44	12.9%
70–74 years	20	5.9%
75–79 years	22	6.5%
80–84 years	21	6.2%
85–89 years	14	4.1%
>90 years	11	3.2%
Race/ethnicity		
White	265	77.9%
Black or African American	35	10.3%
Hispanic or Latino	29	8.5%
Other	11	3.2%

HD, Huntington's disease; MDS, Minimum Data Set; SNF, skilled nursing facility.

Zarowitz BJ, et al, JAMDA 15; 433-428, 2014

**Table 1** Demographics of HD population

Characteristic	SNF	Home	Testing statistic	p Value
No. (%)	228 (7.4)	2,842 (92.6)	NA	NA
Sex, n (%)				
M	84 (37)	1,393 (49)		
F	144 (63)	1,449 (51)	10.79	0.001
Age, y	52	48	3.88	0.0001
Duration, y (SD)	8.6 (5.1)	3.1 (3.4)	14.61	<0.0001

HD = Huntington disease; SNF = skilled nursing facility; NA = not applicable.

**Table 2** Predictors of nursing home placement

Correlate	Hazard ratio	CI
Impaired gait	3.004	1.353–6.668
Impaired tandem	2.546	1.460–4.439
Bradykinesia	1.965	1.083–3.564

Wheelock VL, et al, Neurol 60(6); 998-1001, 2003

# NURSING HOME



Edgemoor Hospital

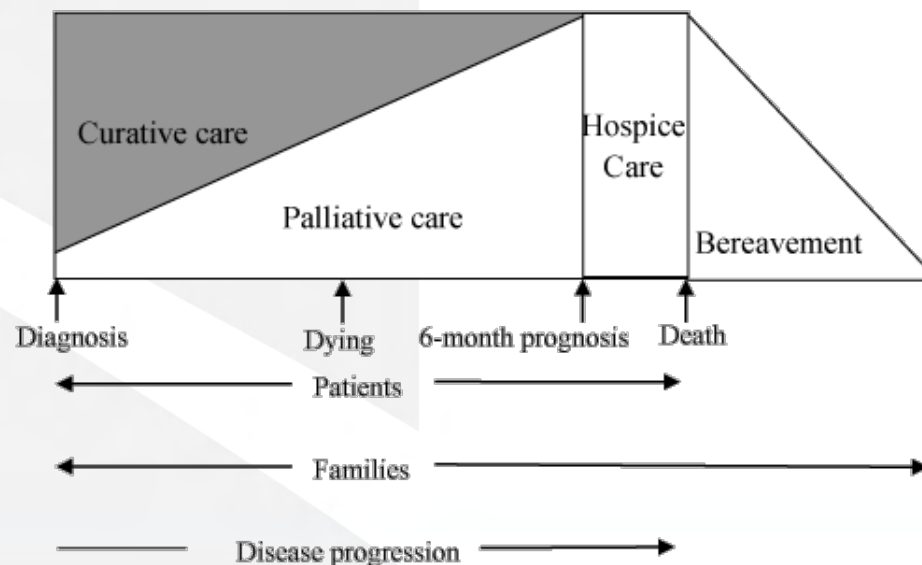
Average age of patients with HD at Edgemoor	51 years
Average age at onset	35 years
Average age at admission (woman and men)	48 years
Average age of men at death	56 years
Average age of women at death	53 years
Average number of diagnosis	6.5
Average number of hospital admissions during SNF stay	1
Average length of stay	7.4 years

*Note.* HD = Huntington disease; SNF = skilled nursing facility.

Dellefield ME, J Neurosci Nurs 43; 186-92, 2011

# PALLIATIVE CARE

...comprehensive, interdisciplinary care focusing on quality of life for patients living with a terminal illness and for their families. Key elements include physical comfort, psychosocial and spiritual support, and provision of coordinated services across various sites of care.”



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Stage V TFC 0 11-26 y	unable	unable	unable	severely impaired	ECF only



## RESEARCH ARTICLE

# Unified Huntington's Disease Rating Scale for Advanced Patients: Validation and Follow-Up Study

Katia Youssov, MD,<sup>1,2,3,4</sup> Guillaume Dolbeau, RI,<sup>1,2,4,5</sup> Patrick Maison, MD, PhD,<sup>2,3,4,6</sup> Marie-Françoise Boissé, MSc,<sup>1</sup>  
Laurent Cleret de Langavant, MD, PhD,<sup>1,2,3,4</sup> Raymund A.C. Roos, MD, PhD,<sup>7</sup> and  
Anne-Catherine Bachoud-Lévi, MD, PhD<sup>1,2,3,4\*</sup>

Movement Disorders, Vol. 28, No. 12, 2013

Advanced HD Functional Capacity Scale					
	Mobility	Feeding	Continence	Communication	Participation
<b>4</b>	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
<b>3</b>	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
<b>2</b>	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
<b>1</b>	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
<b>0</b>	Bed-bound or unable to sit	Most or all nutrition provided by feeding tube	Incontinent and unaware or passive about help	Mute	Non-interactive



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# EXPERIMENTAL THERAPEUTICS

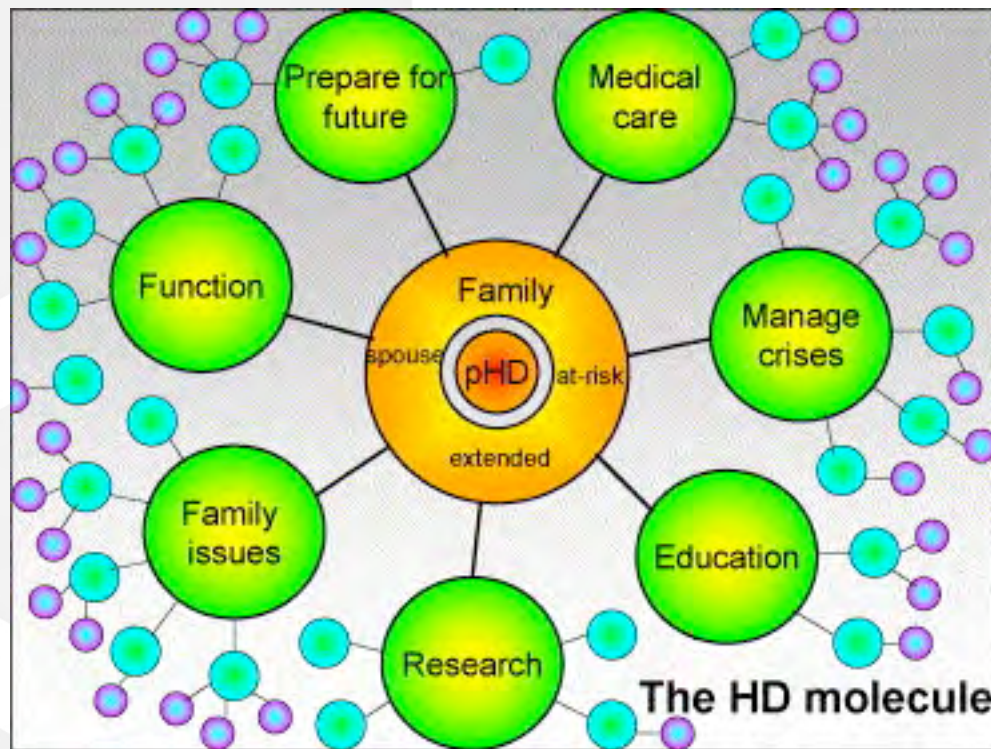
## Medical

- PHAROS
- PREDICT-HD
- COHORT
- TETRA-HD (Tetrabenazine)
- RID-HD (Riluzole)
- TREND-HD (Eicosopentanoic acid)
- HART (ACR16)
- 2CARE (CoEnzyme Q<sub>10</sub>)
- CREST-E (Creatine)

Observational

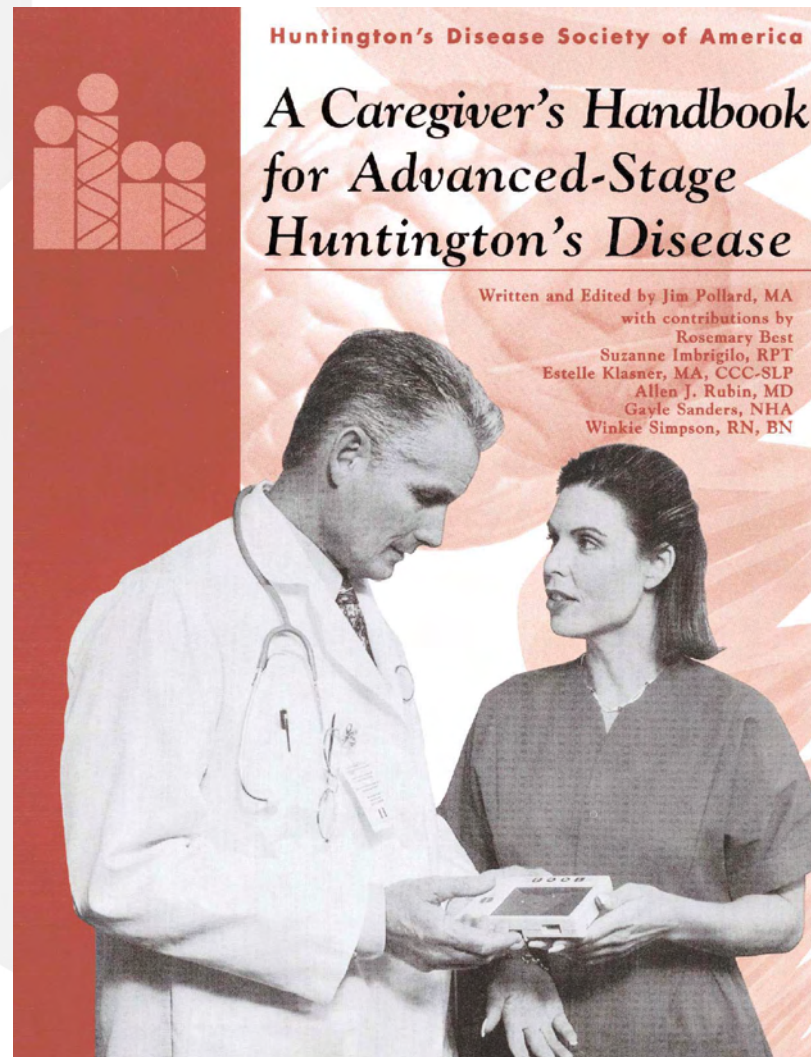
Symptomatic

Protective



Nance, MA, Brain Res Bull, 72(2–3), 30 April 2007, 175–178

# REFERENCES





Huntington's Disease  
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# PALLIATIVE MEDICINE

Specialized area of medicine that addresses care for patients whose diseases are not responsive to curative treatment measures.

# HOSPICE

A program that uses an interdisciplinary team to provide comprehensive palliative care specifically for terminally ill patients.



# UNIFIED HUNTINGTON'S DISEASE RATING SCALE (UHDRS)

17. DIAGNOSIS CONFIDENCE LEVEL

17.

To what degree are you confident that this person meets the operational definition of the unequivocal presence of an otherwise unexplained extrapyramidal movement disorder (e. g., chorea, dystonia, bradykinesia, rigidity) in a subject at risk for HD?

0 = normal (no abnormalities)

1 = non-specific motor abnormalities (less than 50% confidence)

2 = motor abnormalities that may be signs of HD (50% - 89% confidence)

3 = motor abnormalities that are likely signs of HD (90% - 98% confidence)

4 = motor abnormalities that are unequivocal signs of HD ( $\geq 99\%$  confidence)

18. **Motor Examiner**

18.   
-----



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**VI. FUNCTIONAL CAPACITY**

70. OCCUPATION  
0 = unable  
1 = marginal work only  
2 = reduced capacity for usual job  
3 = normal

70.

71. FINANCES  
0 = unable  
1 = major assistance  
2 = slight assistance  
3 = normal

71.

72. DOMESTIC CHORES  
0 = unable  
1 = impaired  
2 = normal

72.

73. ADL  
0 = total care  
1 = gross tasks only  
2 = minimal impairment  
3 = normal

73.

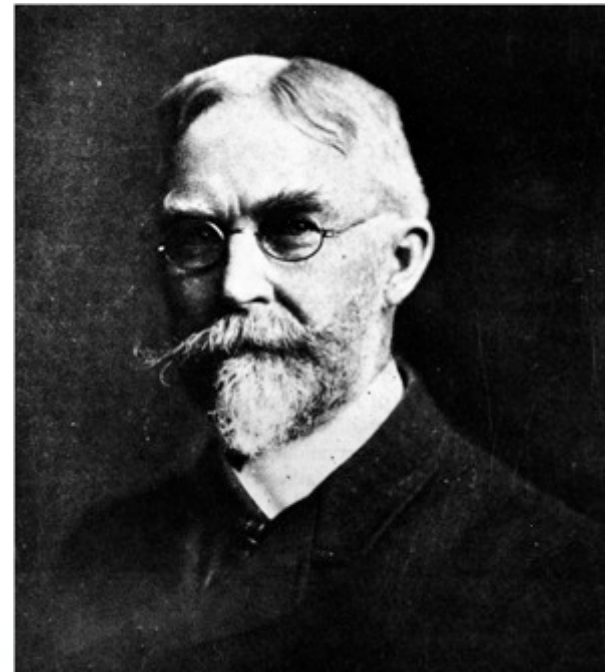
74. CARE LEVEL  
0 = full time skilled nursing  
1 = home or chronic care  
2 = home

74.

# GEORGE HUNTINGTON

Born April 9, 1850, East Hampton, Long Island, NY

Died March 3, 1916, Cairo, NY



# MEDICAL AND SURGICAL REPORTER

## APRIL 1872

“In the history of medicine there are few instances in which a disease has been more accurately, more graphically or more briefly described.”

Sir William Osler  
1908

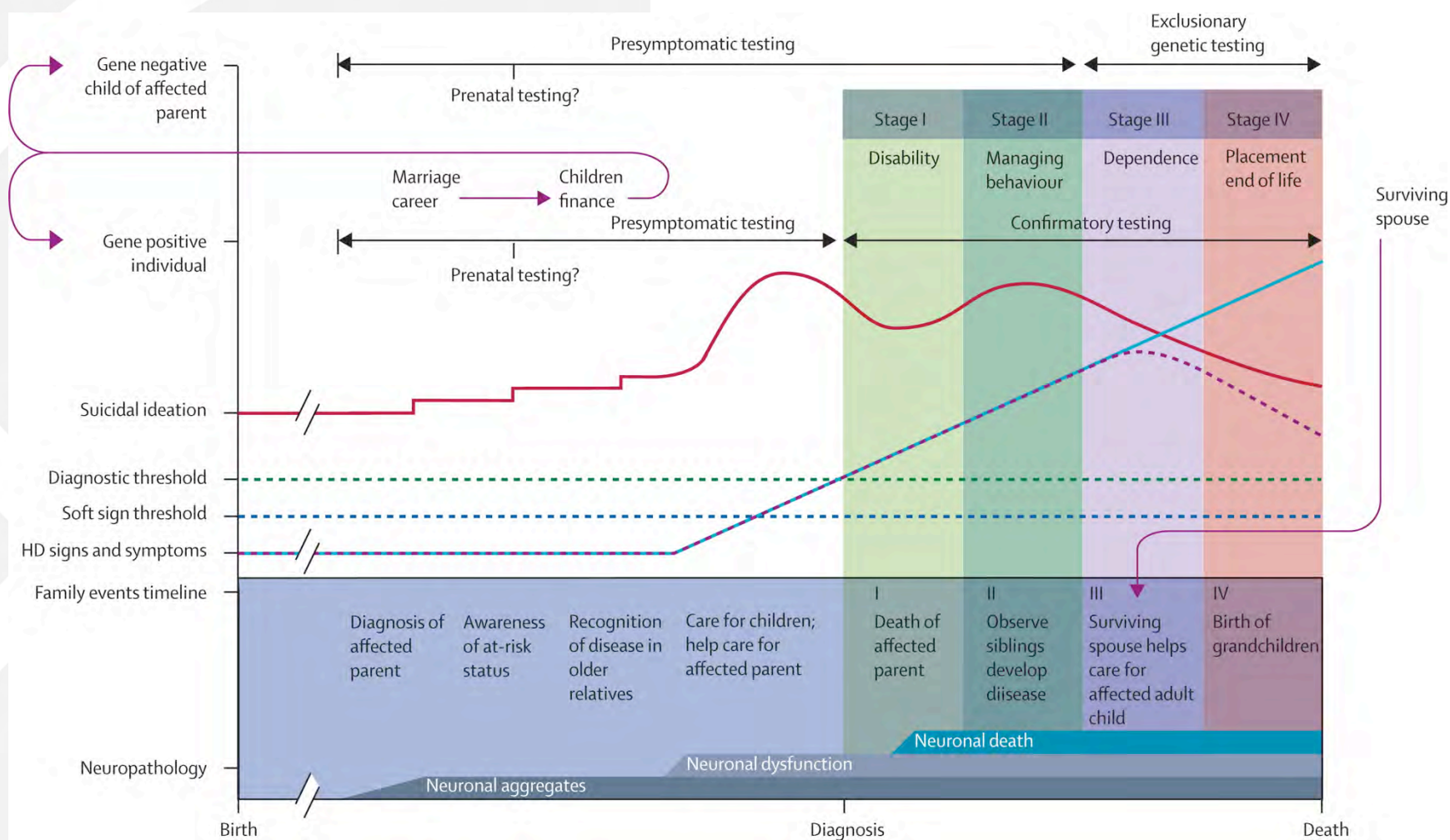


# BEHAVIOR

**Table W-1: Common Progression of Behavioral Stages in HD**

Stage #	Years after onset of illness	Common symptoms
Stage 1	0-8 yrs.	<ul style="list-style-type: none"> <li>• marginal engagement in occupation</li> <li>• independence in all "basic functions"</li> <li>- OR -</li> <li>• maintains engagement in occupation</li> <li>• requires slight assistance in one "basic function"</li> </ul>
Stage 2	3-13 yrs.	<ul style="list-style-type: none"> <li>• unable to work</li> <li>• requires only slight assistance in all "basic functions"</li> <li>- OR -</li> <li>• unable to work</li> <li>• requires major assistance in one "basic function" with only slight assistance in one other "basic function"</li> </ul>
Stage 3	5-16 yrs.	<ul style="list-style-type: none"> <li>• unable to work</li> <li>• requires major assistance with most "basic functions"</li> </ul>
Stage 4	9-21 yrs.	<ul style="list-style-type: none"> <li>• requires major assistance with "basic functions"</li> <li>• may be able to perform some "daily living activities"</li> <li>• care may be provided at home or at a care facility</li> </ul>
Stage 5	11-26 yrs.	<ul style="list-style-type: none"> <li>• requires major assistance with "basic functions"</li> <li>• full-time nursing is usually necessary</li> </ul>

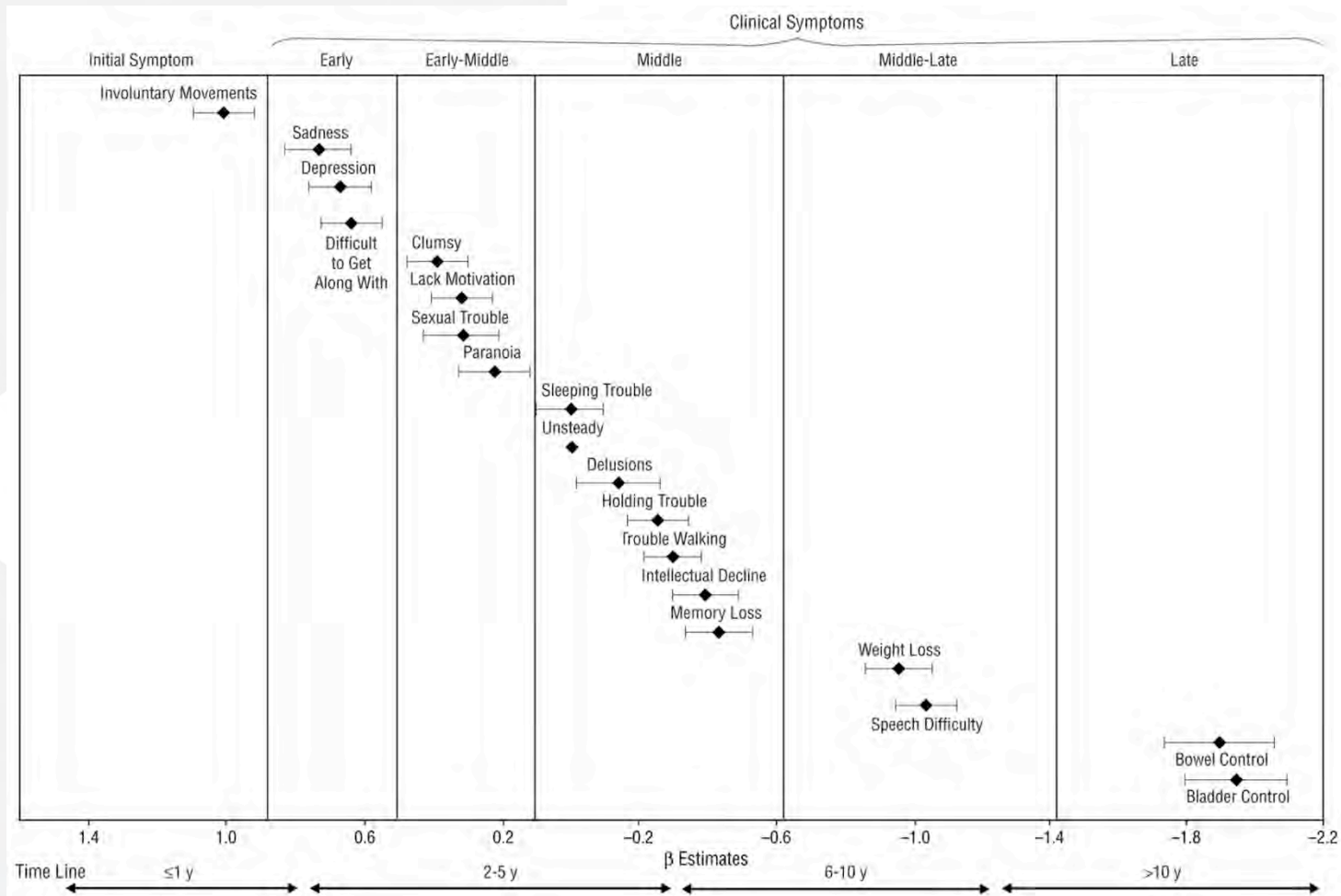




Walker, The Lancet, 369(9557), 20–26 Jan 2007, 218–228



# PROGRESSION



# Treatment of HD

Sedation &  
Mental Slowing

Weight Gain

Tardive  
Dyskinesia

## Involuntary movements

### • Neuroleptics

- Haloperidol (Haldol®)
- Olanzapine (Zyprexa®)

## Mood Disturbance

### • Antidepressants

- Sertraline (Zoloft®)
- Paroxetine (Paxil®)

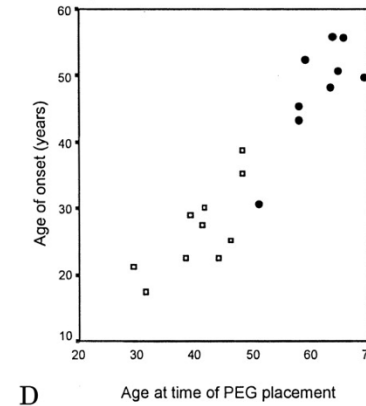
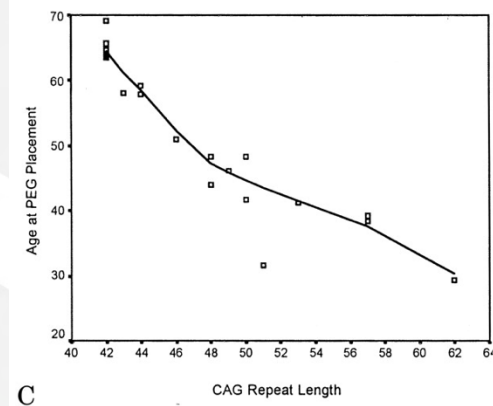
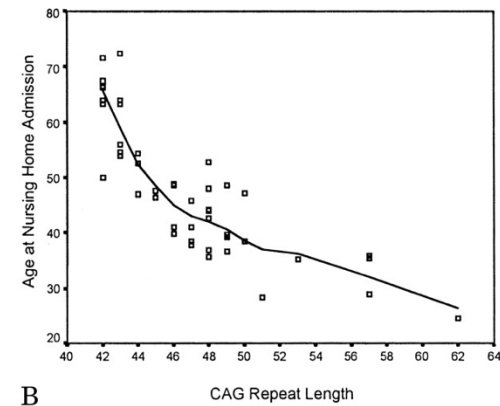
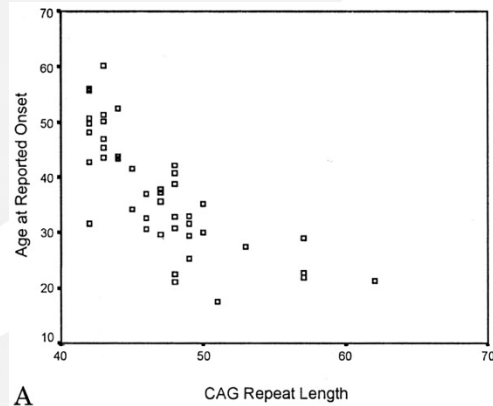
## Paranoia & Psychosis

### • Neuroleptics



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# DYSPHAGIA

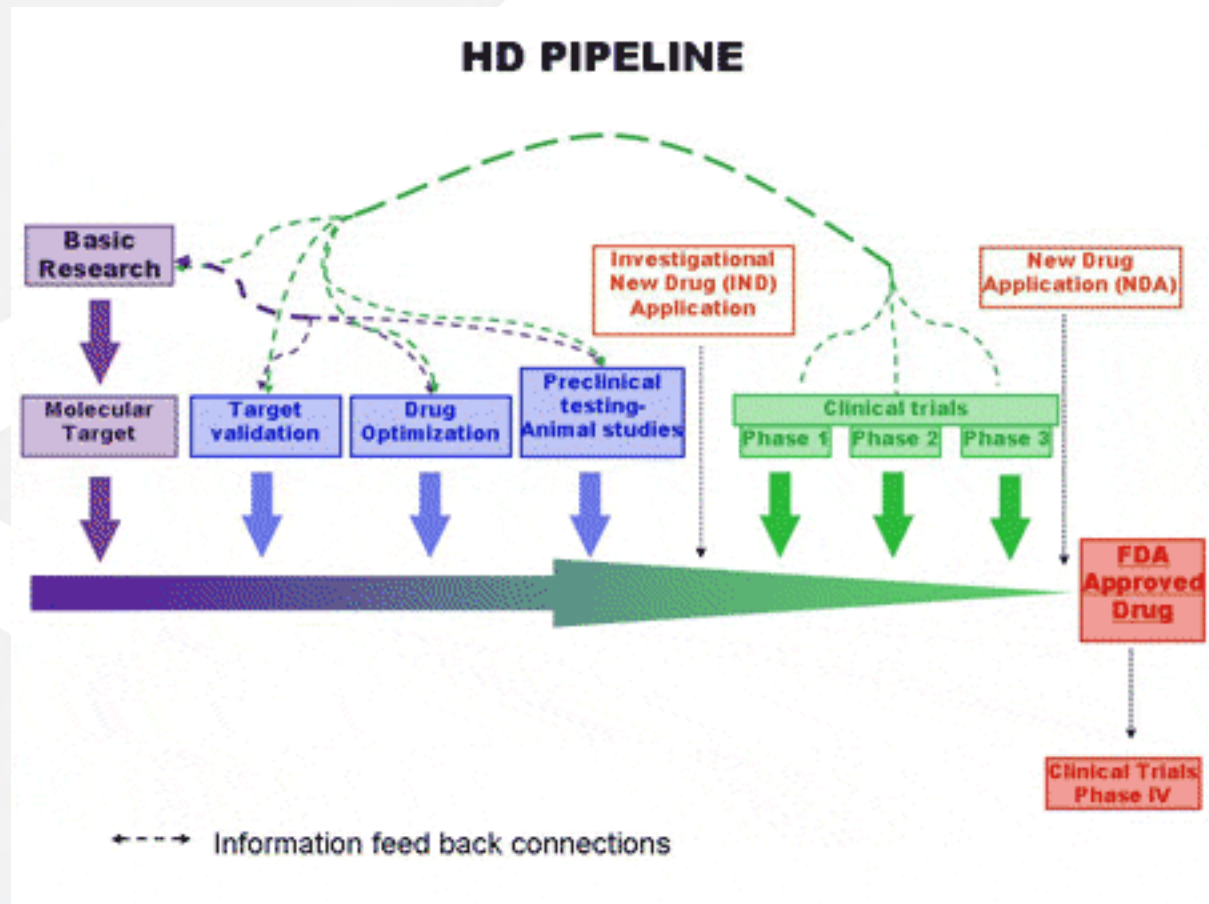


## SEATING

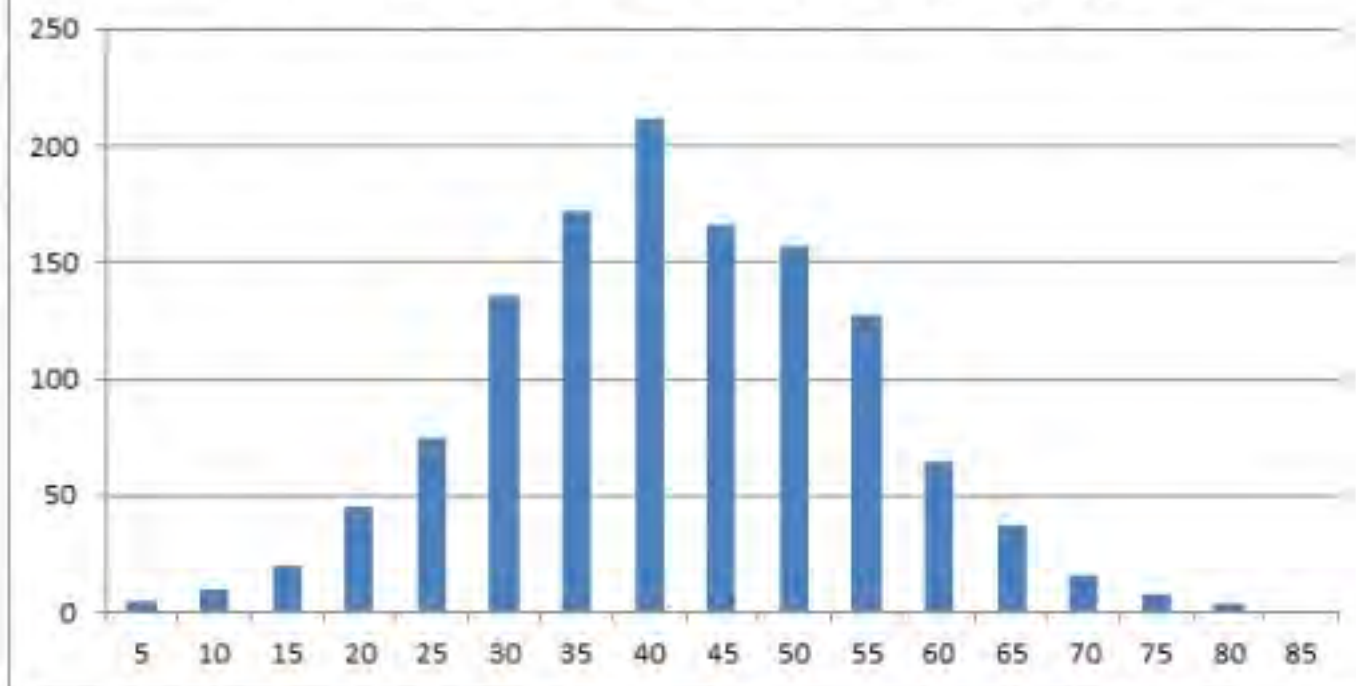


Huntington's Disease  
Society of America

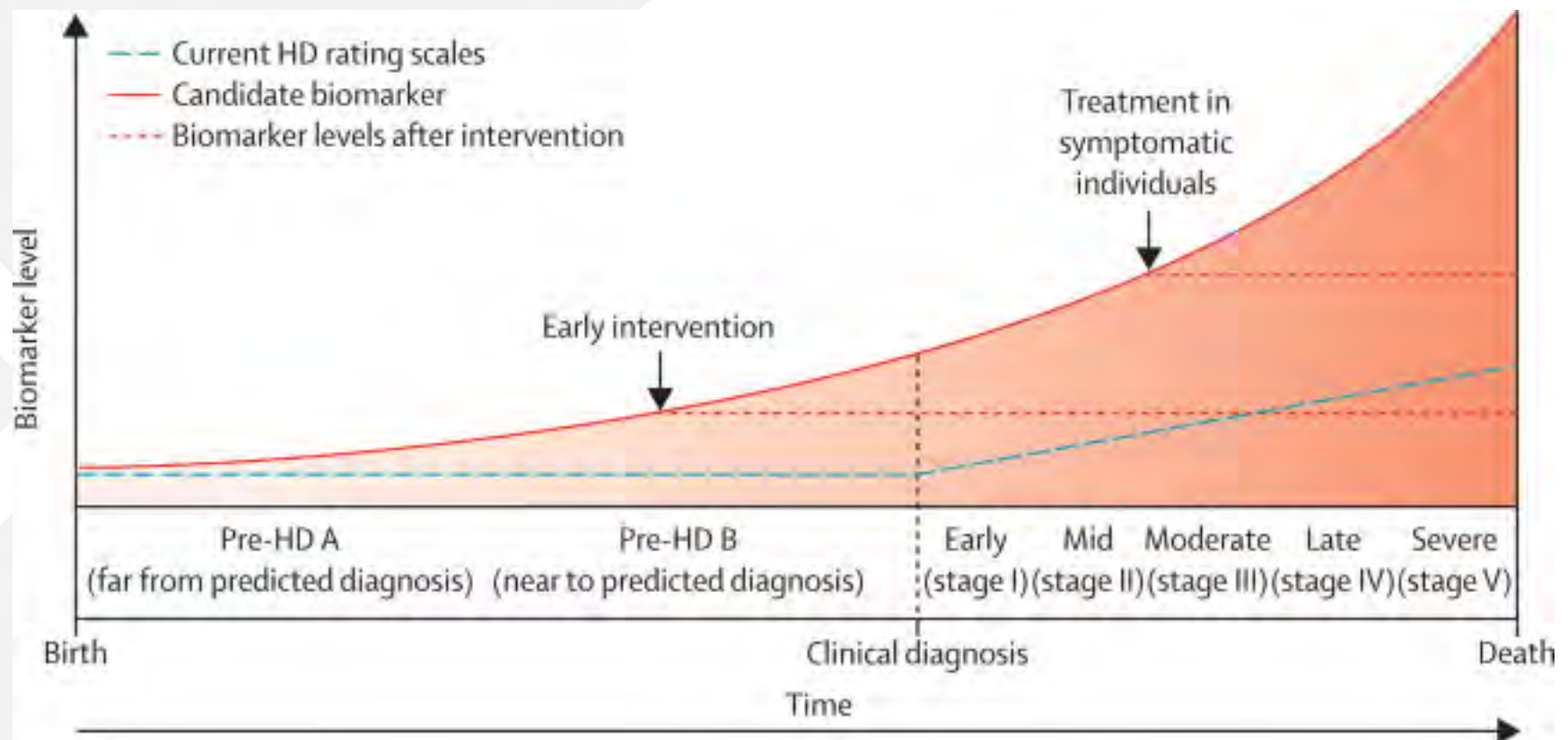
# NOVEL THERAPEUTICS

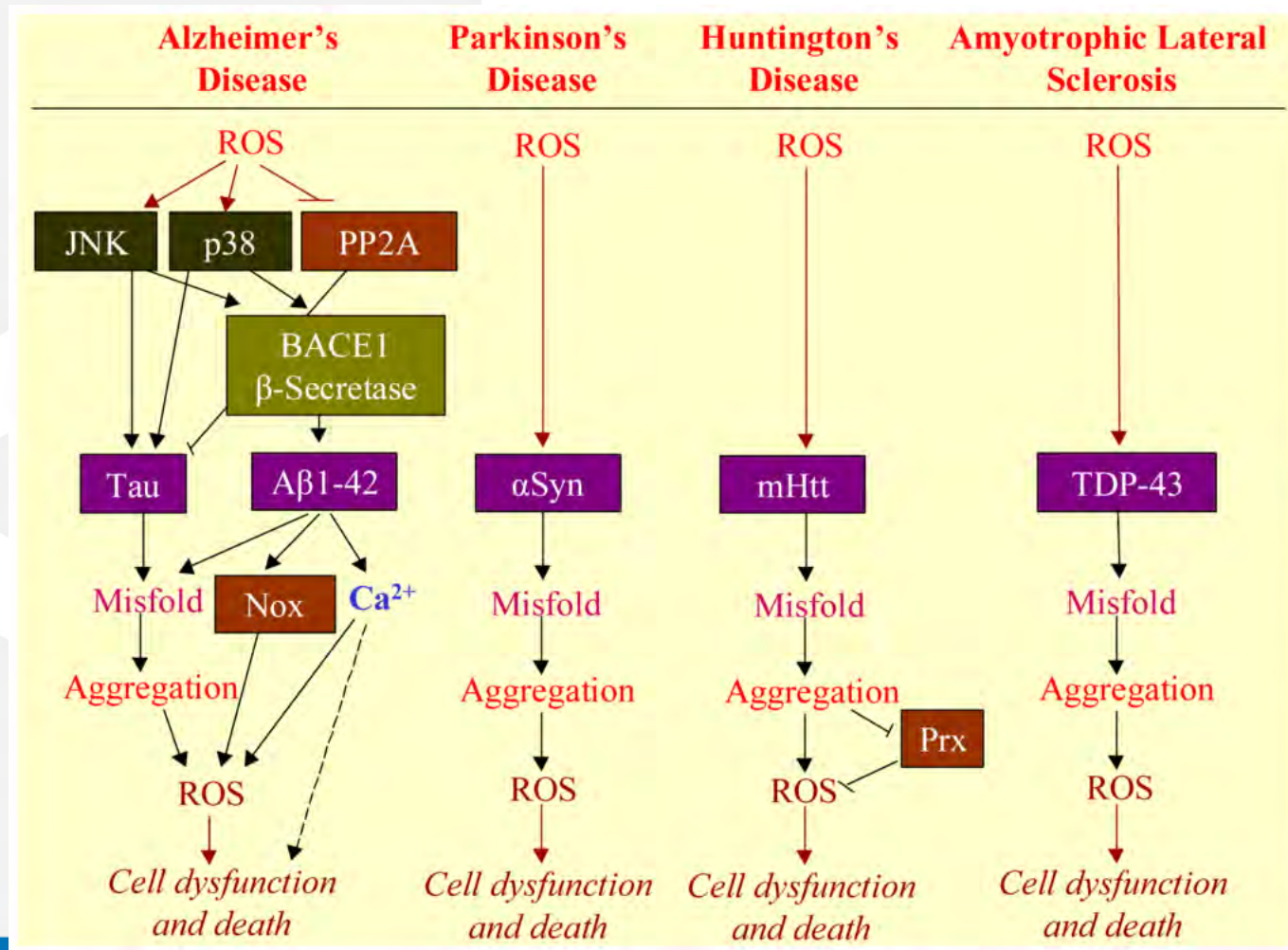


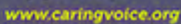
### Huntington's disease occurrence by age











# COGNITION

**Table X-3: Common Progression of Cognitive Stages in HD**

Early Stage	Intermediate Stage	Late Stage
<ul style="list-style-type: none"><li>• Difficulty organizing routine tasks</li><li>• Difficulty coping with changes in routine or new situations</li><li>• Impaired decision-making and attention</li></ul>	<ul style="list-style-type: none"><li>• Increased difficulty working</li><li>• Increased difficulty managing a household</li><li>• Usually capable of carrying out daily living activities such as self-care</li></ul>	<ul style="list-style-type: none"><li>• Can no longer manage daily living activities</li><li>• Often require professional nursing</li><li>• Swallowing and communication difficulties are common</li></ul>



