

Managing Cognitive and Behavioral Symptoms

Robert Y. Moore, MD, PhD, FAAN
Love Family Professor
Department of Neurology
University of Pittsburgh

Director, Huntington's Disease Program
University of Pittsburgh-UPMC

Disclosures

- **Dr Moore has nothing to disclose**
- **However, he does regret not having handouts for his 3 talks. If anyone would like a copy, email a request to his secretary, Joyce Barefoot (barefootj@upmc.edu), and she will send you the slides, or hard copy if you wish.**
- **The talks are**
 - 1. The Genetics of Huntington's Disease**
 - 2. Treatment of Early to Mid-Stage HD**
 - 3. Managing Cognitive and Behavioral Symptoms**

The Motor Manifestations of HD

- **Involuntary movements - chorea**
- **Other motor abnormalities**
- **Bradykinesia – slowness and imprecision of movement**
- **Ataxia – unsteadiness of gait**
- **Eye movement abnormalities – reduced and slowed eye movements**
- **Motor impersistence-tongue**

Behavioral Manifestations of HD - Early

- **Early symptoms**
- In some patients, initial symptoms of HD may include personality changes, such as:
 - Increased irritability
 - A tendency to easily find fault with others
 - Constant complaining
 - Suspiciousness
 - Impulsiveness and lack of self-control
 - Lack of interest in acts that previously provided pleasure (anhedonia)
- Difficulty planning

Behavioral Manifestations of HD – Early to Mid-Disease

Additional behavioral, emotional, or psychiatric disturbances may also become apparent, including:

- Anxiety
- Depression
- Mania
- Obsessive-compulsive behaviors
- Agitation
- Hostile outbursts
- Sleep disturbances
- Increasing social withdrawal
- Severe distortions in thinking may also occur, including false beliefs or delusions, such as fears of persecution. In addition, hallucinations may develop, such as the perception of sounds, sights, or other sensations in the absence of external stimuli.
- **The behavioral manifestations of HD often are exaggerations of the patients normal personality**

Cognitive Decline - Dementia

- **Dementia (from Latin *de-* “apart, away” + *mens-* “mind” - loss of function in multiple domains of cognition)**
 - 1. Orientation**
 - 2. Memory**
 - 3. Executive function**
 - 4. Language**
 - 5. Problem solving**

Mini Mental State Examination (MMSE)

The Mini-Mental State Exam

Patient _____ Examiner _____ Date _____

Maximum Score

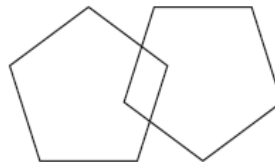
- Orientation**
- 5 () What is the (year) (season) (date) (day) (month)?
- 5 () Where are we (state) (country) (town) (hospital) (floor)?

- Registration**
- 3 () Name 3 objects: 1 second to say each. Then ask the patient
all 3 after you have said them. Give 1 point for each correct answer.
Then repeat them until he/she learns all 3. Count trials and record.
Trials _____

- Attention and Calculation**
- 5 () Serial 7's. 1 point for each correct answer. Stop after 5 answers.
Alternatively spell "world" backward.

- Recall**
- 3 () Ask for the 3 objects repeated above. Give 1 point for each correct answer.

- Language**
- 2 () Name a pencil and watch.
- 1 () Repeat the following "No ifs, ands, or buts"
- 3 () Follow a 3-stage command:
"Take a paper in your hand, fold it in half, and put it on the floor."
- 1 () Read and obey the following: CLOSE YOUR EYES
- 1 () Write a sentence.
- 1 () Copy the design shown.

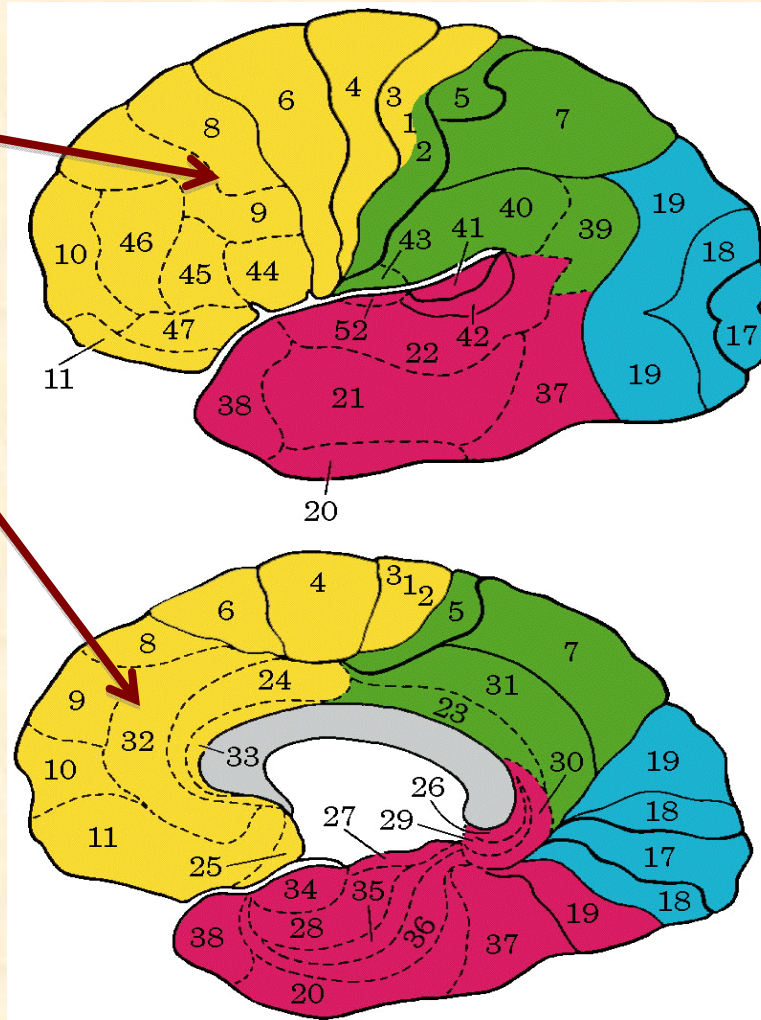


_____ Total Score
ASSESS level of consciousness along a continuum _____
Alert Drowsy Stupor Coma

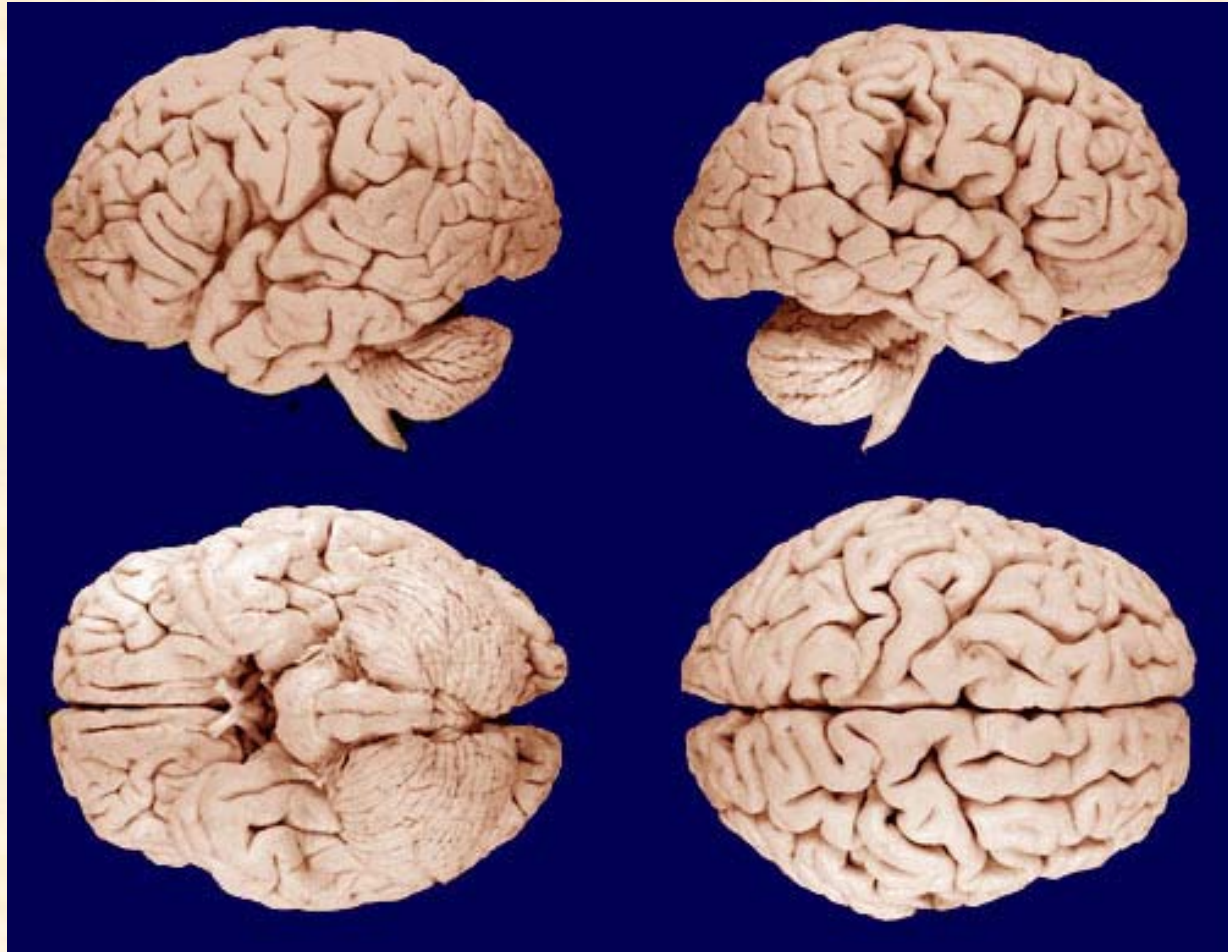
Mechanisms of Behavioral Changes in HD

Frontal Lobes

Frontal lobe



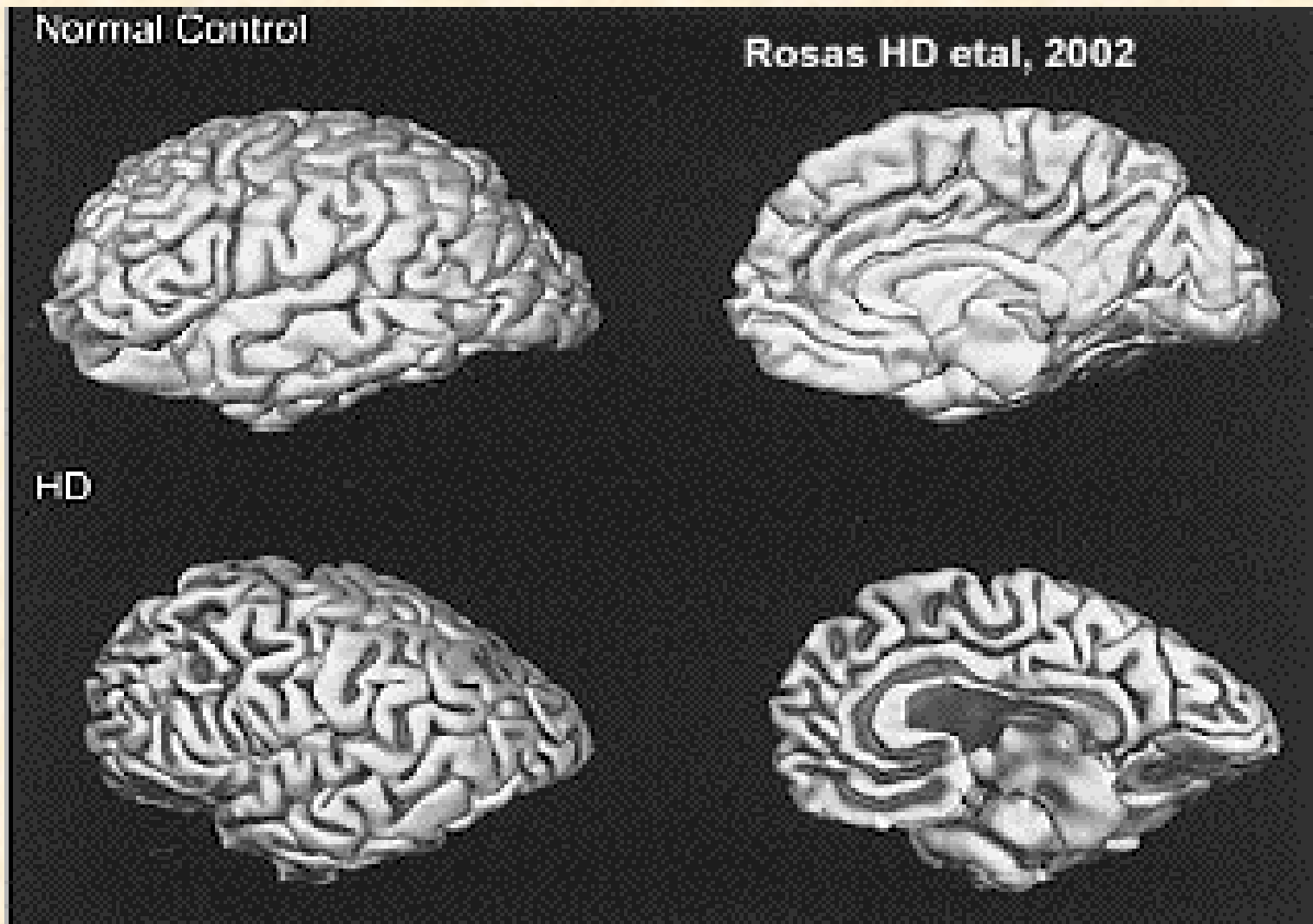
Normal Brain



Brain Imaging

- **MRI images can be reconstructed to show the structure of the brain of living individuals**
- **The following slide shows the cerebral cortex from a group of normal individuals compared to a group of HD patients**

The Frontal Lobes



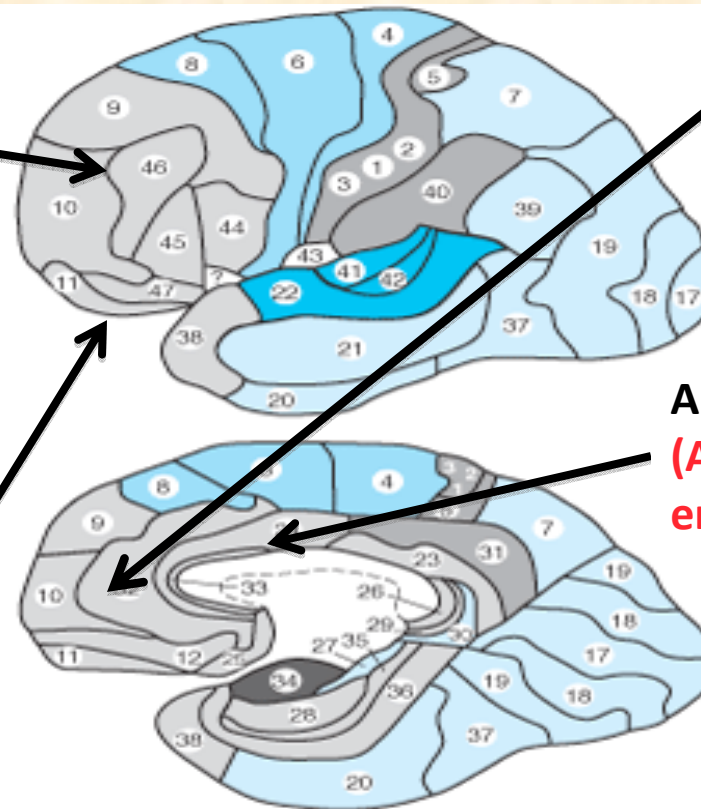
Frontal Lobe Functions

Lateral Prefrontal Cortex
(Working memory, executive function)

Medial Frontal Cortex
(Identity and self)

Orbitofrontal Cortex
(Determining consequences)

Anterior Cingulate Cortex
(Attention, conflict resolution, emotional responsiveness)



light grey	cognition, language, emotion	black	olfaction
dark grey	somato-sensory	medium blue	motor
light blue	vision	white box	???
dark blue	audition		

Depression

A depressive disorder is a syndrome (group of symptoms) that reflects a sad mood exceeding normal sadness or grief. More specifically, the sadness of depression is characterized by a greater intensity and duration and by more severe symptoms and functional disabilities than is normal. The symptoms are characterized not only by negative thoughts, moods, and behaviors, but also by specific changes in bodily functions (for example, irregular eating, sleeping, crying spells, and decreased libido). The functional changes of clinical depression are often called neurovegetative signs. This means that the changes in the brain cause many physical symptoms that result in diminished activity and participation usual activities.

Treatment of Depression

- **Cognitive behavioral therapy (CBT)**
- **Selective serotonin reuptake inhibitors (SSRIs)-fluoxetine, paroxetine, citalopram, sertraline**
- **Selective noradrenaline reuptake inhibitor (SNRI)- duloxetine**

Side effects-nausea, diarrhea, insomnia, headache, dizziness (all usually mild and self-limited), sexual side effects (loss of libido)

Treatment of Depression

- **Bupropion**

Mechanism-unknown

Side effects- agitation, dry mouth, insomnia, headache, nausea-vomiting, constipation, and tremor

Treatment of Behavioral Excesses

(Aggression, OCD, Psychosis, Disinhibition)

- **Behavioral modification – family, friends, reward and punishment**
- **Cognitive behavioral therapy – professional**
- **Medications – atypical antipsychotics**
 - 1. Clozapine (Clozaril)**
 - 2. Risperidone (Risperdal)**
 - 3. Olanzapine (Zyprexa)**
 - 4. Ziprasidone (Geodon)**
 - 5. Aripiprazole (Abilify)**

Side Effects of Atypical Antipsychotics

- **Severe**

1. **Agranulocytosis – clozaril**

2. **Weight gain, diabetes – probably all, olanzepine and risperidone**

3. **Increased mortality in elderly, demented patients**

- **Mild**

1. **Nausea**

2. **Insomnia**

3. **Dizziness**

4. **Headache**

5. **Anxiety**

Treatment of Cognitive Impairment

- **CBT– activity, puzzles, reading**
- **Exercise**
- **Medication**

Memantine (Namenda)

Summary

- **Early HD – look for depression, impairment of judgment, inappropriate behavior, excesses in behavior**
- **Mid HD – signs of dementia, depression, psychotic behavior**

Resources

- **HDSA**
- **Huntington Study Group (HSG)**
- **National Institute of Neurological Diseases and Stroke (NINDS)**
- **Food and Drug Administration (FDA)**
- **RxList (Internet drug index – very informative)**
- **Paying for drugs yourself, check drugstore.com for prices**
- **Last, and not least, your neurologist or regional Huntington's disease clinic**

Everyone Runs Out of Steam from Time to Time



(barefootj@upmc.edu)