



Huntington's Disease  
Society of America

## Cognition and HD

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### Nashville / Vanderbilt University

- Serves the population of Middle Tennessee, parts of Kentucky, Alabama
- <http://www.vanderbilthealth.com/clinicalneurosciences/44384>
- Tennessee HDSA Affiliate



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## OVERVIEW

- *My Assumption:* Cognition is a topic that you have 'thought' about, but may not feel proficient about...
- *Goal:* To give a broad overview of cognitive issues that arise in HD  
Companion talk to this afternoons discussion of therapies
- *Questions we will ask:*
  - (1) Why does this symptom relate to HD?  
Brain-Behavior Relationship
  - (2) How might this manifest in someone with HD?  
Clinical Presentation
  - (3) How can this impact the patient and caregiver relationship?  
Hands on issues

## Topics

- The Affective Domain
- Memory
- Decision Making
- Communication

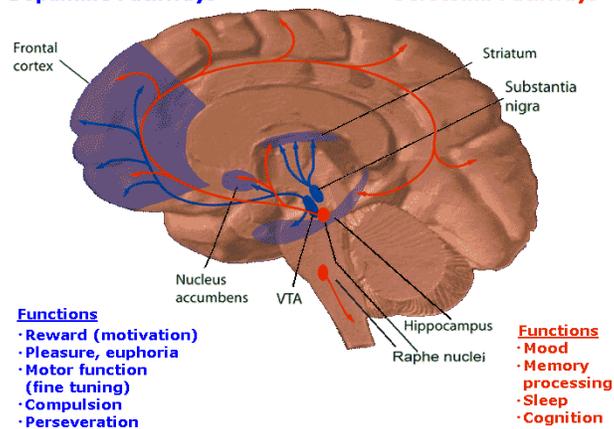
## 2 Minute Brain Tutorial

- Brain is divided into “Cortical” and “Subcortical” Structures
- Neural networks communicate
- “Basal Ganglia” and “Thalamus” integrate networks
- Brain relies on “Neurotransmitters” to enable this communication
- Serotonin (Raphe Nucleus) **Mood/ Anxiety/ Depression**
- Dopamine (Midbrain) **Reward/ Motivation/ Risk Taking**
- Norepinephrine (Locus Ceruleus) **Alertness/ Impulsivity**
- Acetylcholine (Nucleus Basalis of Maynert) **Memory**

Thanks Wikipedia....

### Dopamine Pathways

### Serotonin Pathways



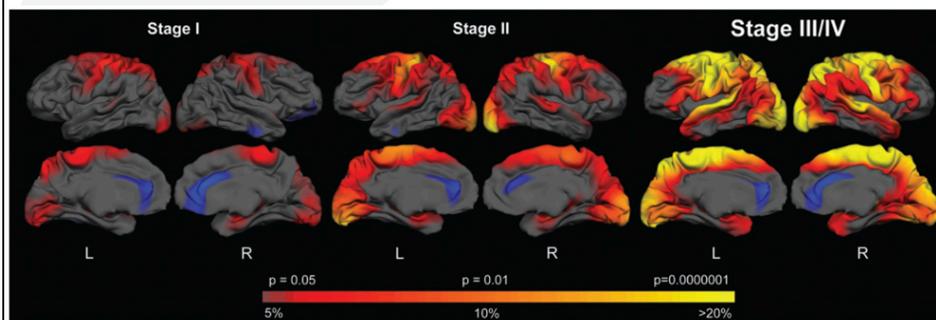
#### Functions

- Reward (motivation)
- Pleasure, euphoria
- Motor function (fine tuning)
- Compulsion
- Perseveration

#### Functions

- Mood
- Memory processing
- Sleep
- Cognition

## How the Brain Changes with HD



### Cerebral cortex and the clinical expression of Huntington's disease: complexity and heterogeneity

*Brain*. 2008 April ; 131(Pt 4): 1057–1068. doi:10.1093/brain/awn025.

## Affective Domain : Cognition, Behavior, and Mood

- Behavioral changes can be most distressing to patients and caregivers
- Specific Areas
  - Depression and Anxiety
  - Obsessive and Compulsive Behaviors
  - Mania and Addictive Behaviors

## Affective Domain : Depression and Anxiety

- Why does this symptom relate to HD?
  - Raphe Nucleus and Dopamine Producing Cells

*Neuroscience* 238 (2013) 71–81

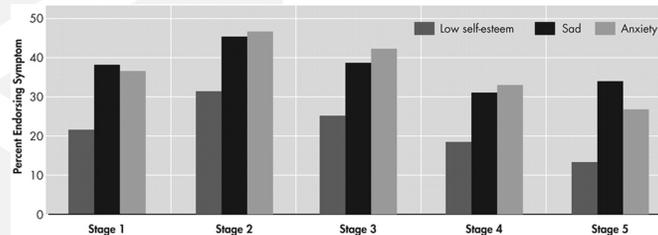
### CHANGES IN BRAINSTEM SEROTONERGIC AND DOPAMINERGIC CELL POPULATIONS IN EXPERIMENTAL AND CLINICAL HUNTINGTON'S DISEASE

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## Depression and Anxiety

- Depression may predate motor symptoms
- It is Common
  - About 40-50% of patients have Depression



From: *Depression and Stages of Huntington's Disease*  
 J Neuropsychiatry Clin Neurosci. 2005;17(4):496-502. doi:10.1176/appi.neuropsych.17.4.496



## Depression and Anxiety

- How might this manifest in someone with HD?
- Depression
  - “discouraged, sad, hopeless, unmotivated, or disinterested in life in general.”
- Anxiety
  - “nervousness, irritability, and problems sleeping and concentrating”
- Suicide Ideations

### DEPRESSION

Depressed Mood  
Lack of Interest in Life  
Changes to Appetite  
Suicidal Ideation

### ANXIETY

Agitated, Worried,  
Not sleeping well  
Trouble Concentrating  
Always Tired

## Depression and Anxiety

- How can this impact the patient and caregiver relationship?

### For the caregiver

- Caregivers experience high levels of stress. They can be frustrated, angry, physically and emotionally drained.
- ~40 to 70% of caregivers have clinically significant symptoms of depression <https://caregiver.org/>

### For the patient

- Impact the relationship with caregiver/ family
- An increase in 'arguments'
- Changes in workplace productivity

## Affective Domain: Obsessive and Compulsive Behaviors

- Why does this symptom relate to HD?
- *Obsessions*: involuntary, uncontrollable thoughts, or impulses that occur repeatedly. "I can't stop them"
- *Compulsions*: behaviors or rituals that one acts out repeatedly. Usually they make obsessions go away.
- *Perseveration*: redundant repetition (cognitive, ideas, motor)
- Frontal cortex (orbitofrontal cortex and anterior cingulate cortex)/ basal ganglia connections control these behaviors

## Obsessive and Compulsive Behaviors

How might this manifest in someone with HD?

- Compulsive hand washing/ worried about contamination
- Stuck to a ritual/ cant sit anywhere but a certain couch

How can this impact the patient and caregiver relationship?

- Difficulty with changes to routine
- Rituals can be frustrating to caregivers

## Affective Domain : Mania and Addictive Behaviors

- Why does this symptom relate to HD?
  - 5-10% of patients develop either bipolar or manic symptoms
  - *Mania*: “feeling high”, irritability, talking fast, restless, engaging in high risk behaviors
  - *Addictive Behaviors*: pathological gambling, compulsive sex/ eating/ buying
  - Frontal (mesial prefrontal cortex) -basal ganglia circuitry control this behavior.

## Mania and Addictive Behaviors

How might this manifest in someone with HD?

- Mania: Mood swings, changes in demeanor, poor judgment
- Addictive and Compulsive behaviors: money, behavior

How can this impact the patient and caregiver relationship?

- Significant stress on caregivers
- Lose large sums of money
- Increase in interpersonal conflict

## Memory

*Working Memory* : “Phone number”

*Procedural Memory* : “How to screw in a lightbulb”

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*Episodic Memory*: “The day we went to the beach with our 5 year old”

*Semantic Memory*: “What is a dog?”

## Memory

- Why does this symptom relate to HD?
- Memory problems impact patients and caregivers significantly
- Manifest in many situations: Lists, multitasks, concentrating, talking
- Can coexist with mood problems
- Memory includes:
  - Encoding new information (a frontal lobe- hippocampal- basal ganglia function)
  - Retrieving information (grossly a hippocampal, frontal/parietal function)

## Memory

How might this manifest in someone with HD?

Biggest issues I encounter in clinic:

- conversational memory
- remembering appointments
- ADLs: when did I last eat? shower?

How can it effect the patient and caregiver?

- Medication management
- Elevate frustration levels over appointments/ conversation
- Increasing reliance on caregiver

## Decision Making

- Very important to maintaining independence, quality of life, ADLs, planning.
- Executive control
- Why does this symptom relate to HD?
- Frontal lobes—basal ganglia function; areas that are altered in HD
- Closely tied to areas involved with mood / behavioral control

## Decision Making

How might this manifest in someone with HD?

- Impulsive behavior: motor (falls) and cognitive (now or later?)
- Managing finances
- Choosing from a menu
- Driving

How can it effect the patient and caregiver?

- When to start shared decision making
- Changes to Independence

## Communication

- “Where the rubber meets the road”

### Specific Areas

- Language
- Reading facial expressions
- Explosive anger (intermittent explosive behavior)

## Language

How might this manifest in someone with HD?

- “Dysarthria”
- Difficulty with naming
- Reduced fluency: simple sentence construction

How can it effect the patient and caregiver?

- Understanding/ Communicating
- Change in hobby
- Relating

## Reading facial expressions



## Reading facial expressions

Why does this symptom relate to HD?

Amygdala, Orbitofrontal cortex are regions that control this process.

How might this manifest in someone with HD?

How can it effect the patient and caregiver?

- Personal interactions become more difficult
- Understanding/ Communicating
- Empathy

## Irritability

Why does this symptom relate to HD?

Amygdala, Orbitofrontal cortex are regions that control this process.

How might this manifest in someone with HD?

How can it effect the patient and caregiver?

- impatience, intolerance, and poorly controlled anger
- ‘Intermittent Explosive Disorder’
- related to impulsivity

## Recap

- Cognitive Changes in HD can happen early
- Cognition is ‘more than memory’ but can include brain processes that guide:
  - Behavior
  - Mood
  - Relationships
  - Communication

## Resources

- [www.nmha.org](http://www.nmha.org) (Mental Health America)
- [www.nami.org](http://www.nami.org) (National Alliance on Mental Illness)
- [www.brainsciencefoundation.org](http://www.brainsciencefoundation.org) (Good section on coping mechanisms)
- [www.aplaceformom.com](http://www.aplaceformom.com) (Great caregiver website)

## Thanks!

