



Denial and Unawareness in Huntington's Disease

Arik Johnson, PsyD

HDSA Center of Excellence at UCLA

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Presenter Disclosure

- Arik Johnson, PsyD
- The following personal financial relationships with commercial interests relevant to this presentation existed during the past 12 months:
- No relationships to disclose

Objectives

- Describe denial and unawareness in Huntington's disease (HD)
- Discuss how denial and unawareness differ and how each impacts people living with HD
- Provide practical recommendations for coping with and addressing denial and unawareness in HD

Huntington's Disease: General Overview

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- Huntington's Disease (HD) is a hereditary neurodegenerative disease caused by an expansion in the huntington gene
- Symptoms of HD fall into three categories:
 - Motor
 - Cognitive
 - Psychiatric

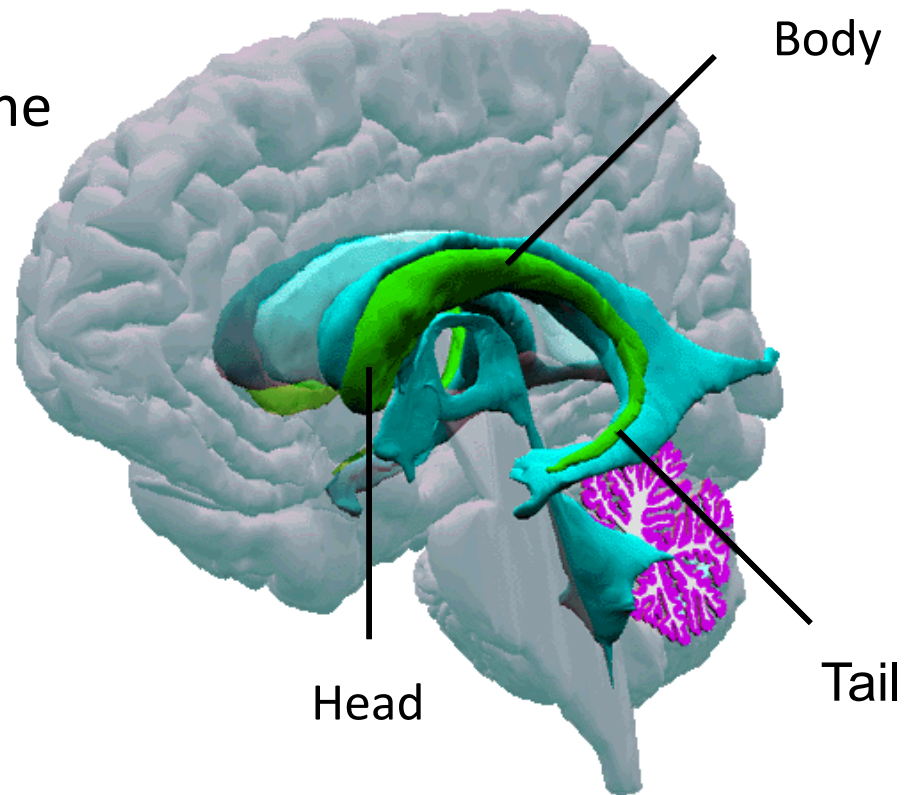
Huntington's Disease: General Overview

- People who have HD generally begin to show symptoms in their mid-thirties to mid-forties
- Symptoms will progress and worsen over time
- There is no way to tell at what pace a person will develop symptoms, how the symptoms will present, or when that individual will die from the disease

Huntington's Disease: General Overview

HD causes changes in the brain itself

- Basal ganglia
 - Caudate nucleus
 - Putamen



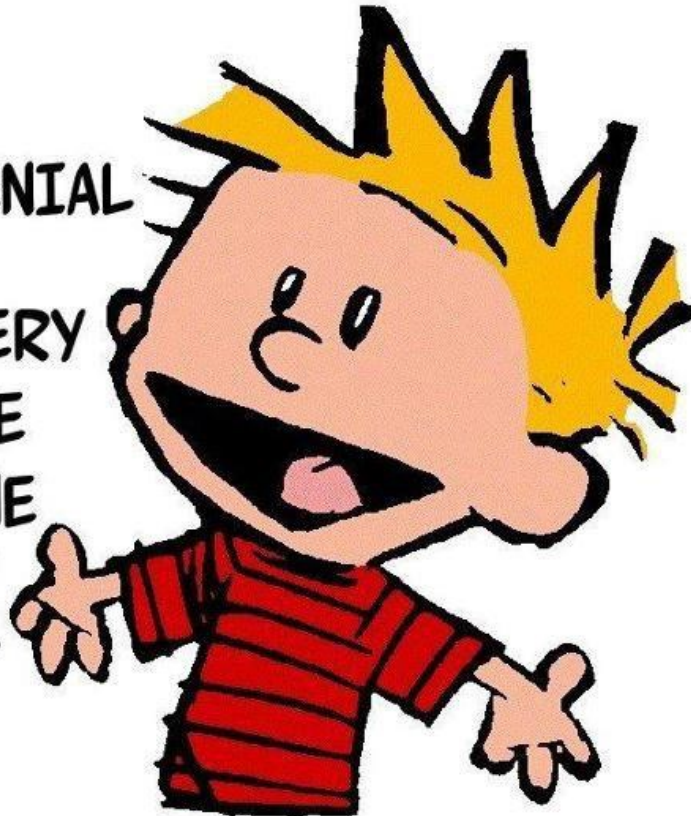
Denial and Unawareness in Huntington's Disease

Denial and Unawareness: Definitions

- Denial
 - A psychological defense mechanism in which confrontation with a personal problem or with reality is avoided by denying the existence of the problem or reality
 - Refusing to admit the truth or reality of something unpleasant
 - www.merriam-webster.com

IT'S NOT DENIAL

I'M JUST VERY
SELECTIVE
ABOUT THE
REALITY
I ACCEPT



Denial and Unawareness: Definitions

- Unawareness
 - The state of being unaware or uninformed
 - NOT having specified facts or feelings actively impressed on the mind
 - www.merriam-webster.com



Denial and Unawareness: Differences

- Denial is an active psychological process
 - Defense mechanism
 - Protects the individual from things that he can not cope with successfully
 - Often there is undeniable evidence that something is true, but the individual continues to deny the information is true
 - Requires a lot of energy

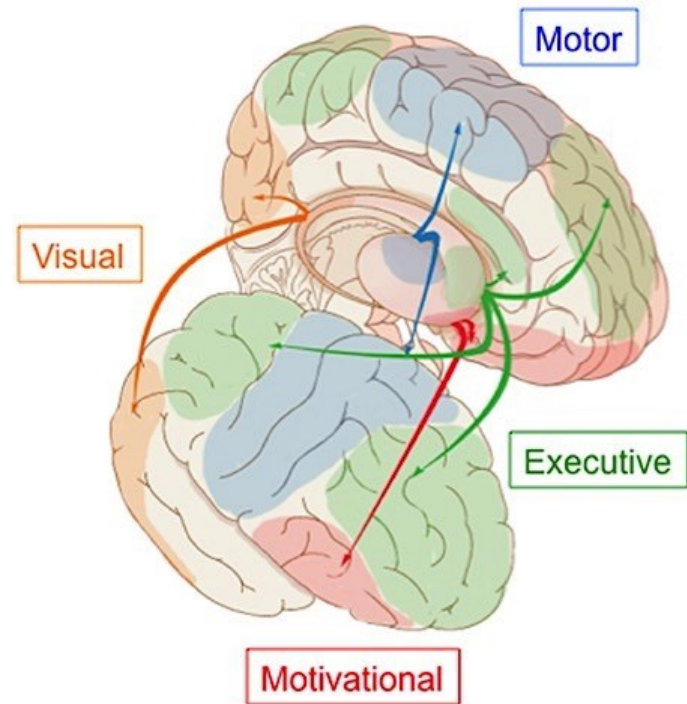


Denial and Unawareness: Differences

- Unawareness is a result of changes in the brain
 - Seen as HD progresses
 - Anosagnosia
 - Lack of self-awareness leading an individual to not recognize her own disability or symptoms of HD

Neuropathology of Unawareness

- Fronto-striatal connections
- Relationship between the caudate nucleus and the frontal cortex
 - Area of the brain responsible for executive functioning



Denial and Unawareness: Examples

- Denial
 - Finding out HD is in the family
 - Refuse to believe the disease exists (“There’s nothing wrong with him”)
 - Minimize the nature of the disease (“It’s just something that mom has”)
 - Not fully understand the disease (“Only boys can inherit it”)
 - Dismiss evidence of symptoms in others (“She can still do everything”)
 - Avoid contact with relatives (“We don’t talk to that side of the family”)

Denial and Unawareness: Examples

- Denial
 - Getting genetic testing results
 - Waiting to receive results (“I don’t want to go in for the results”)
 - Postponing clinical care (“Even though I tested positive, I don’t need to think about it yet”)
 - Not telling family/significant other (“It will only upset them”)
 - Being told that symptoms are starting to manifest
 - Underestimating severity of symptoms (“Even though it takes me a lot longer, I can keep working”)
 - Ignoring clinical recommendations (“I can still drive”)

Denial and Unawareness: Examples

- Unawareness
 - Symptom progression
 - Not recognizing motor symptoms (“I don’t have chorea”)
 - Not acknowledging changes in cognition (“You must have done something to the computer, because it won’t take my password”)
 - Disputing family or clinician input (“You don’t see me at home, I can still cook for myself”)
 - Disconnected from his or her reality (“Why am I here? I don’t even have HD.”)

Denial and Unawareness: A Thought

- What if something that starts as an active choice process turns into a neurological process?

Denial and Unawareness: A Thought

- Denial is a normal response to difficult news
 - Finding out HD is in the family
 - Getting genetic testing results
 - Being told that symptoms are starting to manifest
- Unawareness comes as the disease progresses
 - If an individual is denying that HD is an issue, he or she could eventually lose the ability to recognize that it is an issue

Unawareness

Denial and Unawareness: Recommendations

- There is NO specific medical treatment for denial or unawareness
- There is NO single best way to cope with denial or unawareness
 - Dealing with these symptoms often requires creative thinking and developing interventions for each specific case
 - Interventions are often geared for the people living WITH the person living with HD
 - Family, caregivers, etc.

Denial and Unawareness: Recommendations

- Whenever possible, work with your health care providers to come up with ways to cope with denial and unawareness
 - Team approach
 - Doctor, psychologist, social worker, etc.
 - Family and friends
 - The person with HD
 - Be aware: providers can sometimes feed denial, too
 - Minimizing symptoms (“You’re doing great!”)

Denial and Unawareness: Recommendations

- Treating underlying mood conditions (depression, anxiety) may help
 - Talk about it
 - Talk to your doctors about medication and other interventions
 - Consider therapy for behavioral interventions
- Know when to back off
 - Repeat and retreat

Denial and Unawareness: Recommendations

- Unawareness recommendations
 - Try NOT to interpret inaction or non-compliance as intentional
 - This is *often* a result of unawareness and due to changes in the brain from HD
 - Try to accept that unawareness is a part of HD

Denial and Unawareness: Recommendations

- Unawareness

- Negotiating

- Developing an agreement that outlines the objectives and includes some sort of reward (or consequence)

- “If you go with me to the doctor today, we will stop at McDonald’s on the way home.”

- Contracting

- Putting that agreement in to writing so there is something to refer back to if needed

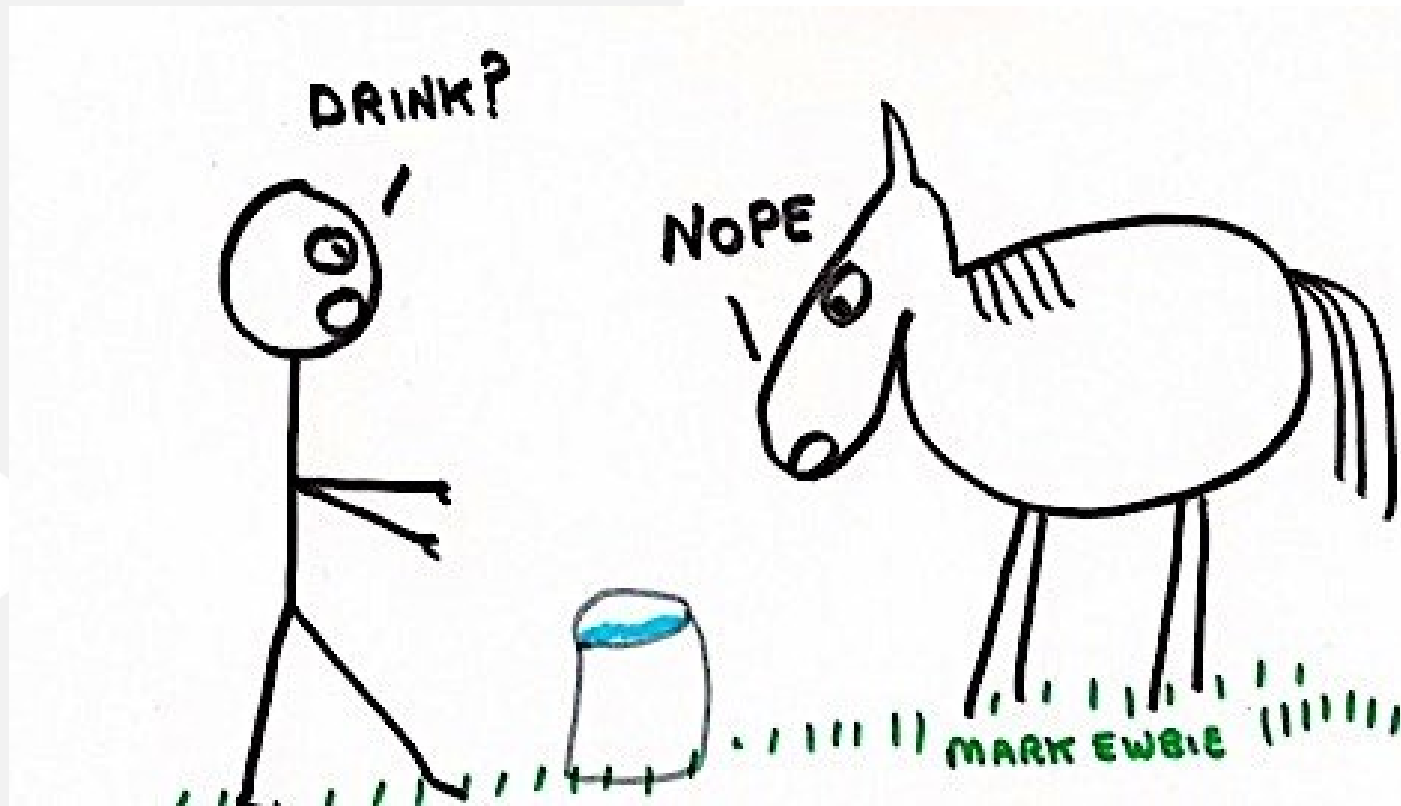
- A woman who volunteers at church agrees to let a friend take her instead of driving herself, with the consequence for driving herself being she would have to give up that position. Her husband sits down and writes this down and the review and sign it together.

Denial and Unawareness: Recommendations

- Denial
 - For the person in denial
 - “When you are ready”
 - Talk about it
 - Consider counseling
 - Work with treatment team
 - Support groups
 - Family
 - Acceptance

Denial and Unawareness: Recommendations

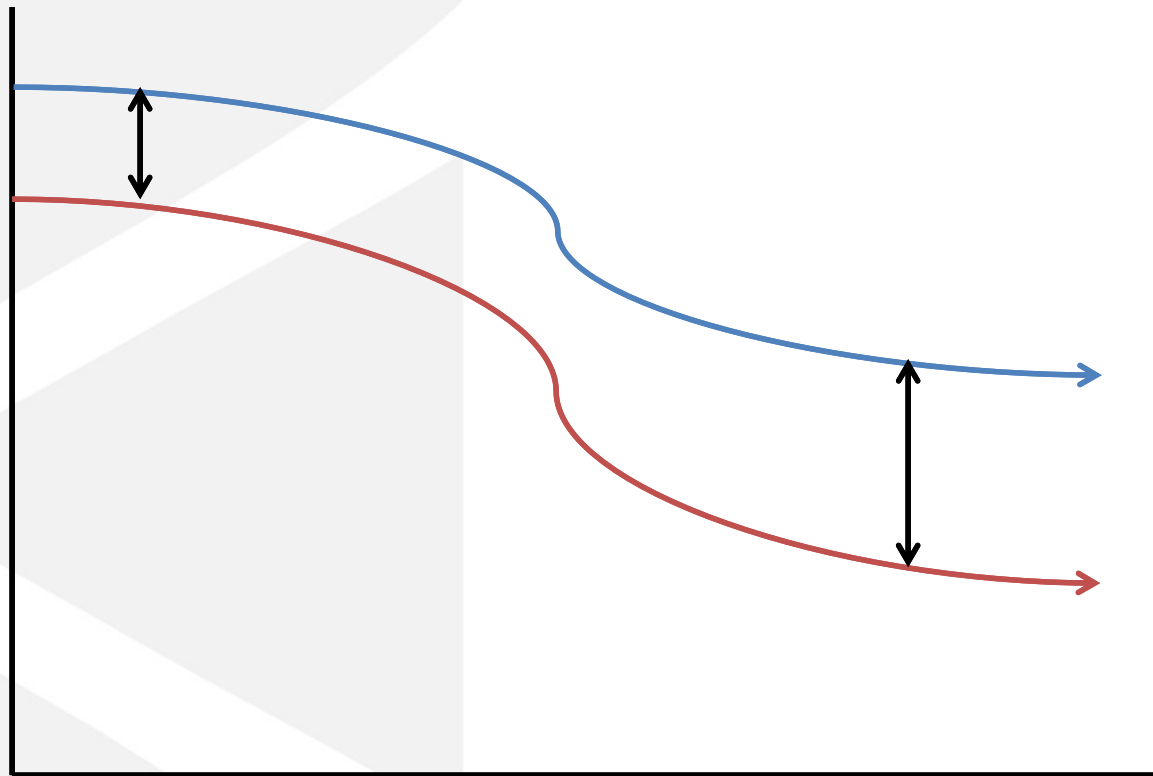
- Denial
 - For the people around the person in denial
 - Understand what “when you are ready” means to her
 - Offer support without overwhelming
 - Encouragement and respect
 - Repeat and retreat
 - Talk about it
 - To someone else for your own peace of mind
 - Acceptance



Denial and Unawareness: Something to Consider

- Starting the process of accepting HD as early as possible might lead to less unawareness as the disease progresses
 - Hypothetical
 - Anecdotal
 - Could it help?

Function



Time

Denial and Unawareness: Something to Consider

- Starting acceptance early
 - Talk about it in the family
 - Consider genetic testing
 - Not for everybody – respect that fact
 - Start a clinical relationship right away
 - Get to know a doctor even before symptoms develop
 - Participate in research

Denial and Unawareness: Something to Consider

- Starting acceptance early
 - Engage in “best practices”
 - Exercise, supplements, therapy, etc.
 - Engage in the community
 - HDSA
 - Chapters and Affiliates
 - Support groups
 - Centers of Excellence
 - Talk about it

Conclusion

- Described denial and unawareness in Huntington's disease (HD)
- Discussed how denial and unawareness differ and the impact on people living with HD
- Provided practical recommendations for dealing with denial and unawareness in HD

This talk should not be the answer
to all of your questions

Hopefully it gives you something to work with and
encourages you to talk to your health care providers
if you are facing these issues

Thank you!

Arik Johnson, PsyD

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June 26, 2015