



Challenging Behaviors in HD: Strategies for Patients and Families

Vicki Wheelock, MD

Lorin Scher, MD

Barbara J. Kocsis, MD

Terry Tempkin, NP

Lisa Mooney, LCSW

HDSA Center of Excellence at UC Davis Medical Center

Huntington's disease (HD) is a *neuropsychiatric* illness, which means that people with HD suffer from both physical symptoms, like chorea, and mental symptoms, such as depression or problems with thinking and memory. Research tells us that psychiatric symptoms in HD often arise years earlier than physical symptoms, and that psychiatric symptoms are frequently the most distressing symptoms for both patients and families to cope with. This article provides an overview of behavioral symptoms common in HD, and suggests strategies for how to best manage them.

Psychiatric problems are extremely common in those suffering from HD; in fact, almost all patients with HD will have at least one psychiatric symptom during their lives, and most patients will have multiple symptoms. Behavioral symptoms in HD are a direct result of changes in the brain caused by the illness. This happens because HD damages important structures and pathways in the brain—and this damage causes the problems with movement, thinking, and behavior. Though the psychiatric problems common in HD can be challenging for patients and families, it's important to remember that these problems are *caused by the illness*, and not by the person suffering from HD. Additionally, it's crucial to keep in mind that people with HD face many losses, including loss of independence, health, and eventually, their lives. These losses burden HD patients with frustration, anger, and grief in addition to any psychiatric symptoms they may have from the illness. Understanding that behavioral issues in HD stem from a combination of brain changes and the effects of profound loss helps to guide coping strategies for patients and families.

Overview of Common Behaviors in HD

Apathy Loss of motivation to attend to responsibilities, basic needs, and things the person used to think were important. *Difficulty getting out of bed or starting the day, neglecting household chores or personal hygiene*

Declining Executive Function Problems with speed of thinking, planning, prioritizing, organizing, concentration, decision-making, flexibility, or creativity. *Poor performance at work, inappropriate behavior, impulsive decision-making.*

Anxiety Intense feelings of inner discomfort, worry, panic, or restlessness. *Frequent worrying about minor or everyday things, fear of losing control or “going crazy,” preoccupation with perceived judgment or scrutiny from others, obsessions/compulsions (see below).*

Depression And Suicide Pattern of low mood and poor energy that can lead to feelings of hopelessness and thoughts about ending life. *Difficulty with concentration, tearful/crying, persistent sadness, irritability, or low energy; changes to appetite and sleep patterns; little or no interest in once pleasurable activities; preoccupation with or frequent thoughts about death or suicide.*
NOTE: Patients with HD are at high risk for depression, impulsivity and suicidality. Unfortunately these symptoms are a part of the illness and should be aggressively treated by your doctor.

Irritability and Disproportionate Anger Often stems from frustration about losses (e.g. abilities, independence) combined with brain changes that decrease the ability to regulate emotions. *Snappy or grouchy tone, screaming, swearing, threatening, slamming doors, hitting walls, pushing, striking or hurting others.*

Perseveration and Obsessive Thoughts and Compulsive Behaviors Fixation or being stuck on one idea or activity. *(ie. ask same question or make the same statement over and over, fixates on specific subject, etc)*
 Recurrent, intrusive thoughts paired with repetitive behaviors that reduce inner discomfort. *Over-concern with germs/contamination, fixation on past insults/injustices (thoughts); repeated hand washing, compulsive eating or drinking (behaviors).*

Psychosis – Delusions And Hallucinations

Delusions are false beliefs, often held with strong conviction. *May be paranoid that someone or something intends to harm the individual.*

Hallucinations may be auditory (sounds or voices), visual (forms, animals, people) or even smells or tastes.

Insight may be preserved (the person recognizes the perception is not real) or absent.

Substance Abuse or Dependence

Includes alcohol or recreational drugs. Abuse or dependence on substances can mask and/or intensify behavior symptoms. *May be used to “self-medicate” from the symptoms of HD. Interferes and/or disrupts daily life, social relationships, work performance, etc.*

Unawareness

Failure to recognize or notice problematic behaviors or the declining ability to function. *Person doesn’t notice worsening performance at work, person fails to recognize they are no longer a safe driver.*

Behavioral Strategies

Apathy and Declining Executive Functioning

Problems with apathy and executive functioning can significantly impact the lives of people with HD and their families. Fortunately, similar strategies are helpful for managing both of these problems. Current medication is not very effective in treating these symptoms.

- Regular routine and a predictable schedule (*e.g. regular meal and bed times, chores completed at same time each day, etc.*)
 - Helps reduce the amount of new information and change the person with HD has to manage.
 - Routine can be soothing and may also reduce anxiety and irritability.
- Cues and prompts
 - Helps the person to remember and pay attention to his/her responsibilities and routine.
 - *Examples include using cell phone alarms, calendars, and gentle verbal prompts to remind individuals with HD that it’s meal or medication time, or that a chore needs completing.*
- Use short sentences that give 1-2 pieces of information at a time
- Offer choices instead of open-ended questions
 - *“Do you want oatmeal or eggs?” instead of “What do you want for breakfast?”*

Irritability, Disproportionate Anger

Sometimes, individuals with HD may experience inappropriate anger that seems out of proportion to the situation at hand. Individuals may lose patience more quickly than they used to, or may not be able to “shrug off” minor irritations. The person’s anger may escalate quickly, and they may yell, slam doors, throw objects, or even hurt others. This can be especially challenging if the person with HD also suffers from unawareness, or the inability to recognize their symptoms or declining functioning.

- Avoid direct confrontation, and adopt a helpful stance when possible.
 - *For example, instead of saying “You can’t drive because you have Huntington’s—your attention and motor skills aren’t good,” try, “I’ll drive you—I was going there today anyway.” This helps the person to maintain a sense of dignity and control, and is less likely to trigger anger.*
- Stop all use of alcohol and recreational drugs.
 - *Though someone using these substances may feel better in the moment, substance use increases the risk of dangerous behavior and is not safe for people with HD.*
- Involve Community/Professional Resources
 - For particularly sensitive issues, such as approaching the person about declining work performance or unsafe driving, involving outside agencies is helpful (e.g. work performance evaluation, driver evaluation through DMV) because it shifts confrontation away from family members.
- Utilize de-escalation techniques
 - Soft voice, kind words, and giving space may help.
 - If the person threatens or uses violence, it is crucial to get away and call for help (e.g. police).
 - Do not attempt to touch or restrain the person yourself. Any HD patient with frequent or severe inappropriate anger should see their HD care team right away, because they may need medications to help reduce anger.
- Limit stress
- Remove any weapons from the home (guns, bullets, knives, etc)
- Maintain a calm, predictable environment when possible

Depression, Anxiety and Suicide

It is common for people with HD to experience depression and/or anxiety, both of which can be treated and managed with medication. The presence of these symptoms can worsen challenging behaviors, therefore recognizing and treating symptoms is critical.

- Medical Evaluation and treatment
 - Medications,
 - counseling or psychotherapy

- Maintaining a consistent and structured routine
 - Use schedules and calendars
 - Simplify routines
 - Allow more time to complete daily tasks
 - Calm environment
- **Suicide:** Be aware that suicide is unfortunately common among those who suffer from HD and depression. Suicidal thoughts and statements in patients with HD *should always be taken seriously and addressed immediately.*
 - If someone with HD expresses intent to harm themselves or has a plan to commit suicide, it is an emergency, and action must be taken.
 - Call a suicide hotline (1-800-273-TALK [8255])
 - 911
 - Or take to nearest Emergency Room for evaluation

Perseveration, Obsessive Thoughts and Compulsive Behaviors

Perseveration and obsessive or compulsive behavior may not cause significant concern for safety or interfere with daily life or activities, however these behaviors are often difficult for loved ones to cope with and manage each day.

- Empathize with their feelings or actions to help them feel understood
- Consistent and structured routine
- Use distraction techniques
 - *change the subject, direct attention to another task, humor, etc*
- Accommodate the behavior (if safe to do so)
 - *i.e. if wanting to always get specific food, buy the food in bulk and store out of sight,*
- Set appropriate limits or boundaries
- Seek medical evaluation for medication

Psychosis: Hallucinations and Delusions

Delusions are false beliefs, often held with strong conviction such as feeling paranoid that someone or something intends to harm them. Hallucinations can be auditory (sounds or voices), visual (forms, animals, people) or even smells or tastes. Insight may be preserved (the person recognizes the perception is not real) or absent. The cause of psychosis may be medical (certain medications, illnesses, infections or traumas) or can be caused by recreational drugs/alcohol. A person experiencing psychosis may be a danger to themselves or other and should be medically evaluated. New onset of delusions and hallucinations is cause for prompt medical evaluation and treatment.

- Medical evaluation
- Remove weapons or other means of harm from the environment
- Avoid use of alcohol, marijuana and other recreational drugs

- Involve police/911 if safety is an issue
- Avoid confrontation that the delusion/hallucination is untrue or unfounded
- Try to work around the beliefs, if safe
 - listen
 - distract or change subject
 - let them express, share belief
- Counseling for loved ones may help identify coping strategies for the family

Alcohol and/or Substance Abuse

Substance abuse is very common in the HD population. Alcohol is a brain toxin and people with HD have less tolerance for the effects of alcohol. Marijuana and other drugs can cause and intensify hallucinations and delusions. Substance use (including alcohol) can cause lack of judgment, poor inhibition, impulsivity, anger outbursts, depression and other psychiatric disturbances.

- Reduce or discontinue alcohol use
- Avoid use of marijuana or recreational drugs
- Understand the signs of abuse or dependence
- Seek treatment as appropriate
- Avoid confrontations while person is under the influence

Unawareness

It is common for persons with HD to experience unawareness of the severity of their symptoms/behaviors. This is often more problematic for the family and caregivers, however can pose a risk to safety as well. Unawareness is the failure to recognize or notice problematic behaviors or the declining ability of function. Unawareness and denial are often used synonymously, but they are different. Denial is psychological inability to cope with distressing circumstances, like loss of a loved one or diagnosis of a terminal disease. Often denial will decrease over time and the person will be able to process the reality. Unawareness is a lack of insight or self-awareness that causes HD persons to be unable to recognize changes in their abilities or evaluate their own behavior. It is caused by the damage and the interruption of circuits in the brain. Denial is thought to be under the control of the individual to “protect” them from reality at that period in time, where by unawareness is not something the HD person can control as they just don’t have any recognition that something is different or changed.

- Offer incentives or rewards for cooperation
 - i.e. offering incentives when the HD person asks others for rides to avoid driving, or staying off ladders (refraining from unsafe tasks) and willingness to allow others to assist despite they don’t feel they need it
- Utilize creative thinking to get HD person to cooperate with a request
 - i.e. disable the vehicle/hide keys to keep the person from driving when unsafe; hire handyman to prevent person from trying to do unsafe tasks themselves.

- Accept that unawareness is a symptom of HD and not the person being non-compliant or purposely uncooperative
 - Try to change your response or reaction to the unawareness
 - Use humor to cope and laugh situations off (if safe to do so)
 - Attend support group for additional ideas and support in dealing with HD

Behavioral Crises or Red Flags

It may not always be clear why a person is behaving a certain way. It is important to be aware of any changes that you or your loved one may be experiencing. Some changes are red flags to indicate that immediate help or intervention may be required.

Seek immediate help for the following:

- Alcohol or recreational drug dependence or abuse
- Anger and aggressive behavior
- Depression with thoughts of suicide, self-harm or injury
- Paranoid thoughts
- “command” hallucinations(i.e, voices) directing the person to harm themselves or others

Crisis Interventions

When you or your loved one is experiencing a behavioral crisis there are some interventions that may be available to evaluate and address the concern safely.

- 911 or Police Involvement
 - 911 and/or police should be called when the safety of any person is in jeopardy
 - Can help de-escalate situations and determine next step to resolve issue
- Emergency Room visits
 - Rapid medical and psychiatric evaluation
 - Come prepared with information about symptoms, behavior issues and length of concern.
 - Bring list of all medications, include any known alcohol or substance use
- Medical Hospitalization
 - Admission into hospital for medical issue needing immediate intervention: medication and/or observation to stabilize or prevent worsening of symptoms.
- Psychiatric Hospitalization
 - Admission directly (voluntary or involuntary) to a facility specializing in mental illness to start/adjust medications and/or observation to stabilize.

Resources/References

- HD Helpline/Social Worker: 916-734-6277
- HD Support Groups: <http://hdsa.org/about-hdsa/support-groups/>
- HDSA Center of Excellence @ UC Davis Medical Center: <https://www.ucdmc.ucdavis.edu/huntingtons/>
- HDSA Northern California Chapter: <http://northernca.hdsa.org/>
- HDSA Publications: <http://hdsa.org/shop/publications/>
 - A Physician's Guide to the Management of HD, 3rd Edition
 - Understanding Behavior in Huntington's Disease: A Guide for Professionals
 - Caregivers Guide to Huntington's Disease
 - Law Enforcement Tool Kit
- Suicide Prevention Helplines:
 - National Suicide Prevention Lifeline: 1-800-273-TALK (8255)

Rev. March 2016

Imooney@ucdavis.edu