

# A Caregiver Guide for HD Families



# A Caregiver Guide for Huntington's Disease Families

## Authors

Nancy R. Downing, PhD, RN, SANE-A, Associate Professor, College of Nursing, Texas A&M Health Science Center

Ann Gaba, EdD, RD, CDN, CDE, Assistant Professor, CUNY School of Public Health at Hunter

Sally L. Gorski, MA, CCC, Manager, Speech-Language Pathology, Hennepin County Medical Center

Deb Kegelmeyer PT, DPT, MS, GCS, Associate Professor, Clinical Health and Rehabilitation Sciences,  
Ohio State University

Anne Kloos, PT, PhD, NCS, Associate Professor, Clinical Health and Rehabilitation Sciences, Ohio State University

K. Michelle Knewstep-Watkins, OTR/L, University of Virginia Health Systems

Anne Leserman, LCSW, HDSA Assistant Director of Community Services

Jessica Marsolek, HDSA Minnesota Chapter Social Worker

Martha A. Nance MD, Director, HDSA Center of Excellence at Hennepin County Medical Center

Karen Tarpata

Janet K. Williams, PhD, RN, FAAN, Professor of Nursing, The University of Iowa

## Special Thanks

Cheryl Erwin, PhD, JD, Director, Center for Ethics, Humanities & Spirituality,  
Texas Tech University Health Sciences Center

Carol Moskowitz, APRN-C

Peg Nopoulos, MD, Professor of Psychiatry, University of Iowa

## Editors

Anne Leserman, LCSW

Deb Lovecky, MS





# Introduction

We hope this guide will provide tools for caregivers to help them at home with their loved one with HD. Professionals in the field who work with persons with HD and their families were instrumental in providing content. In preparation for developing the guide, HDSA surveyed the HD community in order to better understand the issues, behaviors and situations that most impact family caregivers. Our thanks to all who provided their insights and comments.

There is no clear road map on how to care for Your person with HD. We wanted to touch on issues that we know have created challenges for other HD families. We hope that the information included in this guide will inform you about the basics, what things to think about and consider as you incorporate this knowledge into caring for your person with HD.

Most importantly, please know that you are not alone in your struggle. There are many ways to reach out to HD professionals and other caregivers to provide help and strength as you continue on your path.



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# Chapter 1

## Caring for the Caregiver



#### FAMILY NOTE

*I try to separate my feeling about losing my wife an inch at a time and concentrate on taking care of her needs. I am honored to be able to help. It is difficult to stay in this frame of mind but with the help of friends and family I can make it so far.*

## Caring for the Caregiver

Living with HD can mean many things. It can mean that you are symptomatic or that you are at risk for the disease. It can also mean that you are a caregiver for a person with HD. Caregiving is an act of courage, love and sacrifice. As a caregiver, you have chosen to put the wellbeing of a loved one with HD on an equal footing (or even above) your own needs. There is no higher calling than caring for another, but to be effective for the long term, you must also honor and nurture your own body and spirit.

Becoming a caregiver may happen slowly, but it often begins in turmoil, as a response to a diagnosis or a loved one's decline in their ability to meet daily responsibilities. There may be an initial flurry of activity as you try to address the current crises. At that point, questions are typically straight forward and action based – what paperwork needs to be filled out, what does the doctor say, what medications are needed – primarily problems that have immediate solutions. Then, as the initial rush subsides and the depth of commitment required to be a caregiver become apparent, other questions begin to arise – the big questions:

- Why do I have to face this?
- How will I face this?
- Why did things change?
- If everything happens for a reason, what is the purpose of this suffering?

There is no one set of answers to these questions. Finding meaning to life in the presence of illness, suffering and mortality is a profoundly personal journey. The things that may help along the way can include your relationships, your religion/life philosophy, or your appreciation of the eternal in nature, music, and the arts – anything that supports a feeling of connection, value and even joy while you are coping with the difficult and overwhelming tasks of caregiving. It will be an uneven process - two steps forward, one step back, one step off to the side – but worth the effort.





## Working with the Big Changes

Becoming a caregiver is a big shift. Something significant in that relationship has changed forever. It may be that the person you relied on now relies on you, or the “rock” who was steady and reliable has become impulsive and a source of worry, or your easy going family member is now irritable and angry more often than not. There may be a reduction in income or a change in living arrangements.

You may find yourself bending over backwards to adapt to these changing circumstances. You may find yourself adapting to the point where you wonder whether you and your needs even matter anymore. Often caregivers feel that their lives, their wishes and their worth have been swallowed up by the enormity of their loved one’s illness. In the face of chronic disease, caregivers can find themselves without a way to process their own suffering and distress over what has happened to them.

Caregivers may feel tremendous guilt because they are having difficulty coping with all their responsibilities, telling themselves that they have “nothing to complain about” compared to what their loved one is going through. But, by denying the importance of their own experiences and emotions, caregivers can end up withdrawing from life and losing a sense of purpose or meaning to the work they are doing.

## Getting Stuck and Breaking Through

Like all of life, caregiving has its own rhythm of ups and downs, but sometimes it can feel like too much to handle. When responsibilities become overwhelming and you don’t know what course of action to take, you can begin to feel “stuck,” demoralized or clinically depressed. Symptoms of caregiver stress can include trouble sleeping or staying awake, no appetite or too much, no energy or the feeling that you are unable to stop. You may feel helpless, hopeless or incompetent, with emotions ranging from sad or mad to “nothing at all.” You may find yourself self-medicating with alcohol or other substances just to get through the day. Sadly, high levels of stress and unproductive coping responses are common among long term caregivers.

You might ask, why talk about this at all? Aren’t things hard enough? It is essential to talk about the pressures and stresses that caregivers face, not to make anyone feel bad or weak, but to highlight the critical importance of self-care to remain an effective, loving caregiver.

Probably the single most important thing to know in your journey as a caregiver is that you are not alone. Now more than in the past, there is an awareness of the needs of caregivers in our society and with the internet and social networking, there are more and more options for connecting to others who understand your struggles and can help you to build a reserve of personal resiliency.

## Understanding and Managing Uncomfortable Emotions

Big changes can bring up all kinds of feelings. Some of the emotions that arise may not feel good, but know that none of them make you a bad person. As a caregiver, you may feel one way today and another way tomorrow and that is normal.

Many caregivers go through periods of sadness and grief. Grief is not something that waits until a person has passed away, and grieving is not a sign of weakness or selfishness or loss of control; grieving is a way of working through your feelings about what has happened to you and the life you led. Grief lets you experience your feelings about what can never be changed (the past), what has been lost (the present) and what will never be (the future). Grief is not about learning to cope, adjust and survive, though it may help with that. Grieving is a way of acknowledging that the things which have changed had meaning and importance.

Grieving can be very upsetting and it may not be something you should do alone. Do you have a friend, family member or faith leader that you can talk to honestly? Is there anyone who really understands your situation, someone you can talk to on difficult days? Is there someone who makes you feel calm just by being there? If so, set up a time to talk to them, even if it is just on the phone.



If you do not know a person like that now, look for a caregiver support group or grief support group in your community or at a local hospital. It may be an Alzheimer's or Parkinson's disease caregiver support group, but many of the issues will be the same.

The goal is to find people who will understand what you are going through and can be supportive, not judgmental. If feeling of sadness or grief become so strong that they keep you from doing what you need to, it may be wise to make an appointment to talk to a therapist or grief counselor.

Anger is another emotion that can surprise caregivers with its intensity. You may find yourself feeling angry at other members of the family or you may feel angry at the person with HD for their behavior. Anger is often triggered by frustration, exhaustion, or fear. Look for the reasons behind your anger or talk to a friend or counselor. See if there is something underlying your anger that could be changed.

Feeling guilty is also common among caregivers. Guilty that you are angry, guilty that you are well while your person with HD is ill, guilty that you are not superhuman as a caregiver. Guilt and regret change nothing. No one is perfect. Everyone makes mistakes. Your job is to do the best you can at this time and let that be enough.

Loneliness can affect caregivers, even those with big families or communities of faith. You may feel that you have no time for your friends because of your responsibilities or that no one has time for you. You may hesitate to contact a friend because you don't feel cheerful and sociable. You may think that no one understands your problems, but unless you make an effort to reach out to people, you will not know if they understand or not.

If your local circle of support feels weak, look to the Internet. Online support groups, bulletin boards and social networking groups can connect you to people with problems similar to yours that you can communicate with easily. It is possible to connect with an online group even if you only have a few minutes a day. Fight the urge to isolate yourself from others and look for circles of support, either in your community or online. There are people who care about what you have to share.

## Managing Stress

Are there certain aspects of caregiving that "drive you crazy"? Does your life sometimes feel out of control? There is no question that the pressures and demands of caregiving are great, but uncontrolled stress can cause serious physical and mental health problems in caregivers. As difficult as caregiving can be, it is possible to manage your level of stress and finding ways to do that are key to being an effective caregiver over time.

Simply put: stress can be managed two ways: either by changing something in the situation itself or by changing your reaction to the situation.

To change a stressful caregiving situation, you might:

- Prioritize, make lists and establish a daily and weekly routine.
- Identify specific things that trigger outbursts in your person with HD and then work to reduce them.
- Ask for (and accept) help from friends, family or your community of faith. Make a list of simple chores or errands that people could do to lighten your burden.

- Learn about caregiving resources in your community through your local office for the aging or disability services. There may be a local adult day-care or respite center, especially one that treats patients with dementia, which would allow you to schedule some time off for yourself.
- Let your family know if you are financially strapped by caregiving and ask them to contribute to your person with HD's care.
- Be ready to say "no" if asked to take on additional or unnecessary family responsibilities. Know your limits.
- Limit your time with people who offer unwanted advice or judgmental attitudes about your caregiving approach.
- Consider taking a break from your job if you are employed outside the home and you are financially able. Under the federal Family and Medical Leave Act, you may be able to take up to 12 weeks of unpaid leave a year to care for a family member. Your human resources office will have more information.

While there is no easy way to change a situation, you can still change the way you respond to circumstances that arise. It has been said that all you can really control in life is your reaction to it.

- Be kinder to yourself and accept that you are not perfect. There are no perfect caregivers.
- Be kinder to others. Try to forgive those who have not been as helpful and supportive as you would wish. They may be doing the best they can.
- Let go of "ought" and "should" and the need to control how other people behave.
- Try not to be hurt when the person you are caring for is difficult, unreasonable or cruel. It may be the disease talking, not your person with HD.
- If you find that you are filled with anger and resentment, contact a trusted friend or a therapist to help you.

#### **FAMILY NOTE**

*HD does not erase all personality. It hides it, buries it, and alters parts of it, but the person is still there. Their likes, dislikes, sense of humor and full range of emotions are still there. Communication is difficult for a person with HD for a variety of reasons making it seem as though your person with HD is only the disease. It will be even harder for people who don't know your person with HD to see that the person before them is much more than HD. The increased apathy also makes it difficult to recognize the person you love.*

*When I'm having a hard time connecting to my mom I reread the "Changes to Personality" section on a Living with HD article. This helps me to separate the disease from my mom. I also remember what she was like before she became symptomatic. If this doesn't work I think of all the cool cuss words I learned inadvertently from her as a kid and scream them into my pillow.*

## **Making Your Health a Priority**

You cannot care for another if you don't care for yourself. Caregivers often neglect their own health, setting aside things like going to the doctor when they are ill, getting important annual exams such as mammograms, or filling their own prescriptions. Taking care of yourself is not a luxury – it is essential to the wellbeing of the person you are caring for. Eating healthy food, getting sufficient sleep and engaging in physical activity will help you to reduce your stress levels, get more done and perhaps most importantly, maintain the sense that you are an important player in your life story.

Beyond the basics of eating well and getting enough rest, consider making a healthy choice to:

- Do something nice for yourself at least once each day, even if it is just a cup of tea.
- Do something you enjoy. Watch a TV show, work on a hobby, play with your kids or pets.





- Stay in touch with your feelings. Don't judge or dismiss them.
- Keep in touch with your friends. Visit, make a call or send an email.
- Be physically active. Stretch, breathe and get out in nature as often as you can.
- Avoid self-medication with alcohol or other substances.

Taking care of yourself also means seeing yourself in a positive light. Consider all that you accomplish under difficult circumstances. Have you had moments when you were tempted to give up, but didn't? You made the decision to keep going and you should be proud of yourself for persevering. If someone else was taking care of all the things that you do, would you consider that person brave? See yourself as a courageous person, because you are.

Staying healthy is important but some health issues are out of our control. Please attend to your own Advanced Directives about end of life issues

that include how you would like your person with HD to continue to be cared for if you are not able. See 5 Wishes at the end of the chapter for things to consider about your own health concerns.

## Finding Hope and Meaning

It is not unusual to question your beliefs while living with HD. Finding meaning in the face of illness and suffering can be difficult or it can be life changing. Different people have found peace and understanding through practices like reading or listening to uplifting topics, praying, meditating, going to religious services, talking to a spiritual leader or visiting websites that offer a message of hope.

Over the years, researchers have been told by people living with HD that among the losses and sadness, there are positive things to be experienced during the journey with HD. Some people discover that they experience personal growth as a result of the crises they have face, finding:

- Clearer priorities about what is important in life
- A willingness to change things that need changing
- Feelings of self-reliance
- Knowing that there are people they can count on in times of trouble
- A sense of closeness with others
- Knowing that they can handle difficulties
- A willingness to express emotions
- The ability to accept the way things work out
- Appreciation for each day
- Compassion for others
- A willingness to put effort into relationships
- A stronger religious faith
- An acceptance of imperfection
- A willingness to learn
- A gratitude that sees gifts in all reality
- A humanity that relieves the pains of comparison

- A tolerance that accepts difference
- A forgiveness that invites healing.

Please remember that you are not alone. Talk to an HDSA social worker, join a support group, an on-line caregiving group or find an outlet to express yourself.

## **The Five Wishes**

Wishes 1 and 2 are both legal documents. Please check for what legal requirements for an advance directive are required in your state. Wishes 3, 4, and 5 are unique to Five Wishes, in that they address matters of comfort care, spirituality, forgiveness, and final wishes.

### **Wish 1: The Person I Want to Make Care Decisions for Me When I Can't**

This section is an assignment of a health care agent (also called proxy, surrogate, representative, or health care power of attorney). This person makes medical decisions on your behalf if you are unable to speak for yourself.

### **Wish 2: The Kind of Medical Treatment I Want or Don't Want**

This section is a living will—a definition of what life support treatment means to you, and when you would and would not want it.

### **Wish 3: How Comfortable I Want to Be**

This section addresses matters of comfort care—what type of pain management you would like, personal grooming and bathing instructions, and whether you would like to know about options for hospice care, among others.

### **Wish 4: How I Want People to Treat Me**

This section speaks to personal matters, such as whether you would like to be at home, whether you would like someone to pray at your bedside, among others.

### **Wish 5: What I Want My Loved Ones to Know**

This section deals with matters of forgiveness, how you wish to be remembered, and final wishes regarding funeral or memorial plans.

## **Resources**

Caring for the Caregiver <https://www.youtube.com/watch?v=MjcK5dJRCSI>

Advanced Directives

<https://agingwithdignity.org/five-wishes>

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# Chapter 2

## Teen and Young Adult Caregivers



# Teen and Young Adult Caregivers

Are you a teen or young adult helping to take care of a parent or loved one with HD asking some of these questions?

How do I help this person?

What about my life and my plans?

How do I explain this disease to my friends?

If any of these issues apply to you, please keep reading.

## Caregiving by Teens and Young Adults

Many young persons (teens and young adults) living at home provide care to their parent or family member with HD. This is an important responsibility that contributes to the wellbeing of their loved one and to everyone in the family. Some young persons who are caregivers tell us they feel good about helping care for their parent, grandparent, or sibling. One teenager said this: *When this is over, if you don't get it, you'll be a better person in the long haul. You'll have spent your time in hell, so... if you don't get this... then; your life's going to be great because you went through hell already.*

However, being a young caregiver is not easy. As one teen said: *the big question every morning when you wake up is... is it going to be a good day or a bad day?*

When teens and young adult caregivers talk about what is really helpful, the most common responses are that they wanted to spend quality time with their parent or family member, talk with him/her while they can still communicate, and enjoy their relationship with their parent or family member. As one teen said it: *I let my mom tuck me in at night because I didn't know how much longer she could be a mom to me... that helped.*

This chapter describes some of the ways teens and young adult caregivers provide direct care to their parent or family member with HD who lives at home, how the reader can get information about HD and offer some ideas on what might be helpful when carrying out the multiple responsibilities that young persons have. Teen and young adult caregivers face all the issues that older adults do, as described in other chapters in this Guide, plus some unique challenges. Young caregivers are students, employees, and people who want to enjoy relationships with

friends. Each person experiences life as a caregiver in his/her own unique way. This chapter is designed to help the reader recognize the multiple aspects of caregiving that teens and young adults engage in, and offer ideas as to how to manage issues in each of these responsibilities.

### Helping the Person with HD with Daily Needs and Preventing Injury

Caregiving includes helping the parent or family member with HD with daily tasks. This means helping them eat, take a bath, brush their teeth, get dressed, go to the bathroom, walk, prevent falls, prevent injury, take their medications, run errands for them, and talk to their doctor when they are unable to. These are things that young people don't ordinarily do for their parent or family member. However, they are important tasks that help the parent or family member with HD remain safe in their home, and these jobs contribute to the quality of their daily life. They are typical caregiving responsibilities in HD families and may be shared with



the healthy parent. And in some cases, teen or young adult caregivers provide this help on their own, when their healthy parent is away at work, sleeping, or out of the house.

Caregiving for a parent or family member with HD will vary, depending on what works for that individual. What works best may be determined by trial and error, or with help from professional home health care providers. In some cases, teen or young adult caregivers find the solution.

An example of one teen's responsibilities: *Well, I have to feed her, and watch her for my dad so that he can go to work because she can't be alone for more than two hours.* Another young person said: *I worry if I am doing enough.* A third young caregiver explained: *I don't know if it's just because I live with him, but I can pick up on what he's saying. It's usually something totally random, but I can understand it.* The goal in these examples is to protect the person with HD's health and safety.

## Helping the Person with HD Manage Persistent or Upsetting Behaviors

Agitation and irritability are common features of HD, and they are challenging for all caregivers. Teens or young adults may work with family members to limit the person with HD's access to tobacco or alcohol, and in some cases these may be the issues that trigger an angry outburst. If teen or young adult caregivers experience frustration in trying to manage upsetting behaviors in a parent or family member with HD, they are not alone. Teen or young adult caregivers may act as peacemakers to de-escalate the situation for the parent or family member with HD. They may walk away when the parent or family member with HD becomes irritable or angry. This is an especially difficult situation when the person with HD is the young person's parent, and doesn't understand that the teen or young adult is trying to help them. A trusted family member, health care provider, school teacher, counselor, nurse, or coach, or providers at HDSA Centers of Excellence may help the teen or young adult figure out how to manage behaviors by a parent or family member with HD.

The goal is to maintain safety for the young person and for all members of the family. For example: *My dad gets mad really easily.* Another young caregiver described his approach this way: *yeah, relatives who don't see her very much, they don't even know what to do or how to act... like with my grandma, if she gets something in her head and she wants to do it, you have to do it...it's not her fault... and it's not a big deal, but it's hard when they don't understand.* A third explained her situation this way: *to have responsibility, like, you're responsible for yourself but sometimes if you're with your parent, you're really responsible for them too... and make sure that nothing hurts themselves or hurts someone else.*

## Communicating What Works in Caregiving with Others in the Family

It can be really frustrating when a teen or young adult knows what works in caring for the parent or family member with HD, but others in the family disagree. Adults may say they are in charge of making decisions about the parent or family member's care, even when the teen or young adult in the family knows



### Young Persons Checklist for Caregiving

These are topics that young caregivers face. HDSA is here to help. You can access HDSA resources to find help in managing any of these:

1. Helping the person with HD with eating, bathing, walking, sleeping, toileting, preventing injury
2. Helping the person with HD to manage persistent or upsetting behaviors
3. Communicating what works in caregiving with others in the family
4. Taking care of the family
5. Planning for when you won't be living at home
6. Being a student and being a caregiver
7. Taking care of yourself
8. Friends are important

Consider talking to an HDSA social worker, reading the chapters in this Caregiver Guide for HD families and check out the HDSA publication page to read more about HD.

their decisions aren't the ones that make the most sense. Resolving family conflict may be possible when an adult who understands HD (like your family health care provider or HDSA social worker in your region) can help everyone consider how to find what works best in caregiving for the person with HD.

One teen expressed it this way: *I'm kind of stuck, in my family... my dad doesn't know everything that I know... and then he yells at me, and I'm just saying look, I'm trying to help you out here.*

## Taking Care of the Family

Caregiving for a parent or family member with HD often means doing things that help the entire family. Teens or young adults take on responsibilities so that the healthy parent can go to work, or spend less time keeping things going at home. Teens and young adults do the housecleaning, yard work, dish washing and laundry, buy groceries, make meals, and take care of younger brothers and sisters.

This is a lot of responsibility and this means there's less time for school, work, or personal time. For example: *My dad... he doesn't eat, you have to feed him. If he's not cooperating, then Mom has to take care of everything else... so I try to watch over her too. I'll run her errands, clean the house, cook some food, so she takes care of my dad. There's always something that needs to be done.* And another explained: *you're in the middle of something and he needs you right away and you have to drop what you're doing... or else he'll get angry.* Talking with the healthy parent, a trusted adult, or health care provider about how to balance responsibilities at home with other demands may bring up new solutions to sharing these responsibilities while still reserving some personal time.

## Planning for When You Won't Be Living At Home

One of the challenges of being a teen or young adult caregiver is not being able to make decisions about the care of the parent or family member with HD that will be needed when the young person can't be there. When there isn't a responsible adult in the home, teens or young adult caregivers worry about the future and who will take over their tasks when they move out to go to school or start a job. For example: *My main concern, probably is when I go off to college, how's my mom going to get around.... I don't know how she will get her groceries, and what she needs because she doesn't drive anymore. So, just what would she do, because no one would be there all the time?*

## Being a Student or Employee and Being a Caregiver

Many teens or young adults are caregivers and also students in middle school, high school, or college. Some are maintaining a job outside their home. This limits how much time and energy are left over to keep up in school or at work. Caregiving responsibilities can prevent young caregivers from completing homework, getting to school on time, or participating in after school activities. Teachers, school counselors, school nurses, or principals can help create a plan when they know why a young person's responsibilities at home put limits on what they can do to meet expectations at school.

One teen said it this way: *I need my family for sure, but I also need my sports and activities.* Another said: *more time... oh my gosh, work, I live at that place. And I go to work and then go home to my mom. It's like I'm sick of it, I can't do this anymore.* Another young person said it this way: *Just kind of makes it harder, like being the parent & the kid at the same time.... You have 2 lives, one when you're at school & one when you're at home.*

## Taking Care of Yourself

Caregiving takes time and energy from anyone doing it. When young persons in HD families are asked what doesn't help them, the overwhelming response is trying to hold in their emotions. Other problems are not being able to sleep, being too worried to concentrate on school or other topics, worrying about being at risk of HD, and feeling sad.

If a teen or young adult caregiver is at risk for HD, being a caregiver can be an extra emotional burden. Here's what





some have said: *I feel no one knows what I am going through. I need personal time to think more than anything. I'm able to control my emotions better and be more responsible as long as I have a few hours to myself each day.* Another explained: *I have one cousin that I'm really close to, he's a lot older, but he's always been there and ever since I was little, that's who I've told everything to.*

It is a common saying that you can't take care of others if you aren't healthy yourself. And while that may sound pretty simplistic, it has some truth in it. If this sounds like the teen or young adult caregiver in your family, think about how to find a trusted person that young caregiver can talk to in order to help take care of his/her own physical and mental health. This could be someone you know, such as a school nurse or counselor, a doctor, social worker, counselor, or nurse practitioner. It could be someone you don't know, but who can help you, such as members of HDSA's National Youth Alliance or the health care providers at an HDSA Center of Excellence. And use the HDSA helpline (800) 345-HDSA when you need help. There are people at the other end who know about the problems young caregivers experience.

### **Friendships are Important**

Friends are important at all times in our lives, and especially now. When a teen or young adult is a caregiver, it can be especially challenging to find time to be with friends. Friends respond in different ways to the young person's situation and caregiving responsibilities, and young caregivers may have a good idea of how each friend would react. Here is how some did it. *My friends understand, and they know that some nights I just need to be home because my mom's working. I tell her I'll stay home to watch my dad and that gives her comfort. Also, when*



*I go out with my friends, I have it all planned out... There's never a time, like, I can't just leave the house. I need to get it okayed and make sure someone's there to take care of everything.* Another handled friendships this way: *My friends don't know my dad has HD, I see them, I just never ask them over to the house, and a third young person explained: Well, I think it's kind of a benefit for me because it brings out, if something were to happen with him, it brings out, who my true friends are... because they always help me when there's a problem.*

### **Conclusion**

Caregiving is an important and valued responsibility. It requires extraordinary time and energy. Teens and young adults do it in many families. Caregiving will be different for everyone, but some problems are common across HD families. Caregiving involves doing multiple things for the parent or family member while still being a child, student, and friend. Young caregivers can figure out good solutions to some caregiving challenges. But, sometimes added information and support are needed. No solution will work for everyone, and teen or young adult caregivers may need help from several resources.

The main thing to remember is that teens or young adults don't have to do this alone, help is available when you need it. Information, support and problem solving can all be useful to help teen or young adult caregivers, their family, and their parent or family member with HD be safe and as healthy as is possible.



## HDSA Resource list

HDSA helpline number: 800-345-HDSA (4372)

HDSA email contact: [hdsainfo@hdsa.org](mailto:hdsainfo@hdsa.org)

National Youth Alliance <http://hdsa.org/about-hdsa/national-youth-alliance/>

<http://hdsa.org/shop/publications/>

HDSA's National Youth Alliance (NYA) motivates youth to get involved in their local HDSA Chapters, Affiliates, and Support Groups in efforts through education, fundraising, advocacy and awareness for Huntington's disease. The NYA is a collection of children, teens and young adults from across the country ages 9-29 that are impacted by Huntington's. The NYA is dedicated to the vision of being the last generation with Huntington's disease and as an essential part of the future of HDSA. Contact the NYA at <http://hdsa.org/about-hdsa/national-youth-alliance/>

Caregiver Corner Webinar- Talking with Kids about HD <http://hdsa.org/living-with-hd/caregivers-corner-webinars/talking-with-kids-about-hd/>



# Chapter 3

## Social Services in Late Stage Huntington's Disease



# Social Services

*Do you have questions about long-term care, finding local resources, applying for disability or getting support by phone or in a support group?*

*Consider talking with a social worker.*

Much has been written about providing a team to assist in the care of a person with HD. The care team might include the following professionals: a Nutritionist might need to address diet issues along with a Speech-Language Pathologist to evaluate the swallowing mechanism of the person who may have excessive coughing or drooling that could lead to aspiration. Typically a Neurologist saw the person to address movement issues. A Psychiatrist or a Psychiatric Nurse Practitioner may assist with medications for irritability or anger and help caregivers for “as needed medications.” An Occupational Therapist and Physical Therapist can assist the person with HD as they transition from being ambulatory to needing a walker to needing a wheelchair. These professionals can also offer suggestions about adequate bedding, proper toileting and safety in the environment where the person resides. And Social Work services may be needed at various times for referrals, HD information, emotional support and assistance to families.

Many of the caregivers filling out the HDSA survey that have helped to guide this publication looked to social workers for guidance on a multitude of issues facing HD families.

## Why a Social Worker?

Social workers are licensed professionals trained to advocate, support and assist in navigating resources with their clients. City or County Human Services/Social Services offices, hospitals, nursing homes, correctional facilities and school settings employ social workers. A county social worker may help navigate available resources that can assist in the care of a person with HD. Hospital social workers assist in both inpatient and outpatient care arenas. They may help to ensure a safe transition home, provide support when applying for resources or refer to other community resources. Nursing home (also called Nursing and Rehabilitation Center or Skilled Nursing Facility) social workers support the resident and family’s mental health needs, support behavior plans devised by the facility, discuss the prospective resident’s advance directives, and provide assistance during transitional times. Correctional facility social workers assist in transition planning and coordinating mental health needs. School social workers are able to aid students during family changes and provide a listening ear to children.

## Finding a Social Worker

HDSA chapters and affiliates and HDSA Centers of Excellence hire social workers to support families affected by HD. Use the locator button at [www.hdsa.org](http://www.hdsa.org) to find a social worker in your area. The interactive map will assist in finding the social worker closest to you with HD expertise. The national HDSA office may also assist in providing resources and support. Call 800 345-HDSA.

## Support Groups

Many of those that responded to the caregiver survey found value in being a part of a support group. HDSA support groups across the country are led primarily by social workers.





#### FAMILY NOTE

*The support group meeting we attend monthly has kept me from becoming overly bitter and angry. Support groups were helpful to know what to watch for in the future. You are not alone. Support groups can be helpful to both the caregiver and the person with HD.*

## Financial and Insurance Benefits

Social Security Disability Insurance (SSDI) pays benefits to those with a disability when they are no longer able to work. SSDI is based on the number of years you have worked and paid taxes into the federal system. The number of years needed to qualify for benefits will depend on your age.

It is not necessary to hire an attorney or a non-attorney Social Security Representative or Advocate to assist with a disability claim but it is important to provide thorough documentation to the Social Security Administration about how the person with HD is totally disabled from the disease. This often includes reports from multiple sources with information on how the person is disabled. Employers and caregivers can also provide information along with the medical reports as part of the disability claim. Consider watching the Caregiver Corner webinars on disability for more assistance.

Supplemental Security Income (SSI) pays benefits based on financial need. Please use the Social Security Administration website ([www.ssa.gov](http://www.ssa.gov)) as a helpful tool to apply for benefits or to assess eligibility.

Webinars on disability are on the HDSA website under Caregiver Corner.

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/>

Applying for Disability

Disability Strategies 2

Applying for Disability Benefits for HD: The SSA Perspective

Caring Voice Coalition ([www.caringvoice.org](http://www.caringvoice.org)) is an organization that assists patients at no cost with applications for Social Security Disability Insurance (SSDI) and Social Security Income (SSI). Caring Voice advocates help to coordinate supporting evidence, assess eligibility, submit claims for benefits and draft arguments.

Long Term Care (LTC) Insurance policies can assist in future caregiving needs such as in-home care, assisted living, nursing home care and respite care. Long-term care insurance should be considered prior to diagnosis as plans can cover home health services as well as nursing home care. Coverage for persons diagnosed with HD is not guaranteed under the law. For more information on this topic, the American Association for Long Term Care Insurance website might be helpful. <http://www.aaltci.org/>

## Paying for Services

Private pay or paying out of pocket is always an option as all providers accept private pay funds. If a family has income or assets to pay for care services there may be more choice of providers.

Private insurance through an employer or private policy may cover some services. Check with the insurance company to understand what specific benefits are offered and how to access them.

Medicare is health insurance for those 65 and older OR for those who are permanently disabled.




Medicaid or State Health Insurance may be available to those that are disabled who meet certain income or resource limits.

Some states have Waiver programs that use new and existing ways to deliver and pay for health care services. Check to see if your state has a Medicaid Home & Community-Based Waiver to pay for home care through your local County Health and Human Services agency. Ask the local office about other services in your area. Use the Internet to search for specific resources in your county or region. Ask the HDSA social worker in your region about programs specific to your state.

## Ancillary Services

Ancillary services include a wide range of health care services to support the work of a primary physician. Specifically, therapeutic ancillary services include home health, Physical Therapy, Occupational Therapy and Speech-Language Pathology. There were recent changes to the Medicare benefit for these services that eliminates improvement as required to obtain continued service. This is an important distinction for persons with HD that can benefit from services but may not necessarily show improvement.

## State/County Health and Human Services



Aging and Disability can be found by searching for Area Agency on Aging (that supports individuals who are disabled as well as aging.) [www.n4a.org](http://www.n4a.org)

U.S. Department of Health and Human Services, Administration on Community Living  
<http://www.aoa.gov/> and also to find local resources <http://www.eldercare.gov/>

Home care options might be available in most communities. Companion services are paid professionals who provide companionship, supervision, light housekeeping, errands and meal preparation for individuals who are living in their homes. Home care services are paid professionals who are able to provide similar services as a companion but also are able to provide needed “hands on” caretaking (toileting, bathing, medication administration.) Home care services are able to assist a family in the goal of having an individual live at home safely.

Respite services offer a short break or vacation from caregiving that can help to rejuvenate and alleviate burnout, common for many caregivers. Respite can be provided at the individual’s home, nursing home or group home. Services could be family or friends who stay with the person with HD or a professional respite organization, elder care referral service or community or senior center offering older adult “daycare.” Respite services are at times paid for through insurance which includes insurance through the Affordable Care Act or private pay. Length of time depends on insurance or assets available to support this service. Respite resources can include Adult Day Health Care Programs, In-Home Assistance and Community/Senior Centers. There may also be friends and family who can be organized to assist with caregiving and household responsibilities to offer the caregiver some type of break.

Some areas have home care companion services or volunteer companion services that may provide respite for caregivers for a few hours in a day, to allow caregivers to run errands, work or do an activity for themselves.

Ask your local area on aging or ARCH National Respite Network for resources in your community.

**[Archrespite.org/respitelocator](http://Archrespite.org/respitelocator)**

### FAMILY NOTE

*My mother was admitted to a PACE program with a shared aide program to help keep her living independently in her apartment. While she had to move to one of the apartment buildings covered by the program, the aides and one nurse are in the building and check on her twice a day, manage her medications and serve one hot meal 5 days a week.*

Is there a PACE program in your community? PACE (Program of All-inclusive Care for the Elderly) is a Medicare and Medicaid program that helps people meet their health care needs in the community. Medicare or Medicaid will provide more information about this program or others that might be similar. <https://www.medicare.gov/your-medicare-costs/help-paying-costs/pace/pace.html>



## Long Term Care

If you are considering placement in a long-term care facility, please allow time to learn about facilities and the process of placement. Finding a facility that has available beds and is knowledgeable about HD may be difficult. Many facilities are reluctant to work with persons with HD. Careful thought into what is best for the person and what will make them the happiest is the most important consideration when you are faced with options.

For more information about finding resources, consider this US Department of Health and Human Services website. [www.longtermcare.gov](http://www.longtermcare.gov)

Please use the Long-Term Care Guide for further information on this topic and what things to consider. <http://hdsa.org/wp-content/uploads/2015/05/long-term-care.pdf>

Also there is a Caregiver Corner webinar on this topic.

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/long-term-care-hd/>

## Legal Resources

If you are looking for legal advice as you prepare advanced directives or plan financially for long-term care, consider talking with an elder care attorney. This legal specialty encompasses estate planning and looks at persons with disability as well as the elderly. <https://www.naela.org>

## Advanced Care Planning

Please review HDSA's Family Guide series on Advance Directives to help answer questions on guardianship, power of attorney, special needs trusts, etc.

[http://hdsa.org/wp-content/uploads/2015/03/HDSA\\_Family-Guide-Advance-Directives-for-HD.pdf](http://hdsa.org/wp-content/uploads/2015/03/HDSA_Family-Guide-Advance-Directives-for-HD.pdf)

It is very important for a person with HD to share their wishes about their future care early in the disease. It is critical to have a trusted person identified as the financial and healthcare power of attorney (POA) or guardian. This is especially important prior to entering a care setting. There may be difficulty finding someone who is willing or able to take on the task so the care setting may have to enlist a professional guardian to be the person's advocate. This professional assistance with finances or major decisions may prevent excess strain in family or friendship relationships.

## Hospice/Palliative Care

Hospice services, available in many settings such as a home, hospital or nursing home, create a comfortable environment for a person in the terminal stages of HD. Recurrent hospital stays and severe weight loss may be signs of late stage HD. Hospice staff help the person with HD and the family to experience death as an expected, comfortable, and peaceful outcome of the disease.



## Palliative Services

Palliative care can offer a different viewpoint from traditional medical perspectives by focusing on alleviating suffering of individuals and families. Palliative care offers a program where conscious decisions made about care include a thorough assessment of a person and his or her family's quality of life as the disease progresses.

## How to Educate Others About HD Using Available Resources

If you live far from an HDSA Center of Excellence or major metropolitan city, it may be difficult to find professionals who are knowledgeable about HD. But the professionals you encounter can become knowledgeable and use their professional gifts to offer care to a person with HD. The responsibility may fall to the caregiver to educate the professionals caring for your person with HD in order to better understand the individual differences that occur with every person with HD.

HDSA has several publications (some free, some at minimal cost) to assist in understanding and learning more about HD. *A Physician's Guide to the Management of Huntington's Disease* is a wonderful resource that has applicable information for patients, caregivers and care providers, in addition to physicians. It is free to download from the HDSA.org website. <http://hdsa.org/shop/publications/>.

HD101 is a course for physicians to learn more about HD and also get free Continuing Education Units. The course can be found <http://hdsa.org/for-healthcare-professionals/>

You can educate local emergency personnel by making contact with your local police or fire departments and by sharing the HDSA Law Enforcement tool kit. HDSA social workers are specifically trained to provide in-services about HD to care providers and interested groups. Contact an HDSA social worker to inquire about their ability to educate a group or agency that would benefit from knowing more about HD. If an HDSA social worker is not able to provide an in-person training, webinars and conference call trainings are available.

On the HDSA website, free webinars and previous educational presentations from past national conventions are available on a wide variety of topics.

Caregiver Corner webinars <http://hdsa.org/living-with-hd/caregivers-corner-webinars/>

Caregiver Interventions

Cognitive Strategies Part 1 and Part 2

Managing Psychiatric Symptoms

Crisis Situations and De-escalation Strategies

### FAMILY NOTE

*Don't keep the family history a secret. Advocate for your person with HD. Never give up.*

## HD Family Dynamics

For an HD social worker, it is important to move beyond the basic explanation of a triad of symptoms challenging a person with HD. The impact on a family with a disease that is inherited and generational can be tremendous. The person with HD has likely seen other family members progress and die from HD complications. They may feel guilty about the children they have that are at risk for the disease. They may recall family members who committed suicide or spent time in jail or were in psychiatric facilities for treatment with unknown diagnoses. And because



of their illness and their poor judgment or inability to monitor their reactions to events, they may have alienated many or all of the people who once cared for them. All of this will impact the person with HD as they become more dependent on their caregivers.

Some family members will be supportive and want to be involved in the care of their loved one. Others may not be able to face the progression of HD in their loved one or cope with their own “at risk” status.

For all these reasons, the family dynamics are complex and unique in an HD family. It is important to know who in the family is best able to be supportive at this time, knowing that adult children or siblings may not be able to help because of their own fears of developing symptoms, or because they are developing symptoms. For some families it can feel like “looking into a mirror” of what may very well be in their own future. The best allies may be extended family, who may be slightly more removed from the situation and possibly more able to be helpful. Knowing about the progression of symptoms in HD is important for the person in your care — but also being aware of possible early cognitive or behavioral changes in those at risk may assist you in working with your family. Families can become “overly involved” in the care of their person with HD and delay adjustment for both. Adult children and well spouses may feel that they have to keep a family member at home or they will think they have “broken” a promise that they would never place their family member in a nursing home. Placing a loved one with HD in a nursing home setting may be required for the safety of the loved one or care provider. Family members should not put themselves at risk of physical or emotional harm to care for their loved one.

Due to the rare nature of HD, families may be much more knowledgeable about the disease than many health care providers. Families affected by HD become used to being the “expert” on HD but alternatively may grow tired and resentful of this burden.

## Cognitive Concerns

Please also see the HDSA publication *Understanding Behavior in HD* for more detail about these issues.

<http://hdsa.org/wp-content/uploads/2015/03/Understanding-Behavior.pdf>

Anosagnosia is a term meaning unawareness of symptoms. This is not a psychologically-based denial (although this can sometimes be seen, as well), but an “organic denial” that results from deterioration of brain cells. It often seems odd to notice a person with HD who has obvious motor symptoms like gait imbalance or severe twitching of hands, feet and mouth, who repeatedly denies that they have trouble walking. They may even deny the existence of HD in themselves or of the disease in their family.

Trying to convince someone with a brain disease of an issue can be frustrating and likely futile. It is important to work around issues like needing to bathe or change clothes rather than a direct approach that may cause lashing out or a verbal attack. Sometimes taking more time with a person with HD to accommodate their particular likes and dislikes can, in the end, create more calm and compliance than pushing an issue and having the person overreact. The response to the person’s overreaction will likely be more time consuming and could put the caregiver at risk if the person lashes out.

## Assessing Behavioral Problems

Behavioral disturbances are often the reason that a family will request long term care or as a referral from a hospital after a short-term inpatient stay (often psychiatric). Learn to be a good advocate for the person with HD to provide needed information to other professional caregivers about how best to work around the psychiatric and cognitive issues that the person with HD might present. Consider talking with an HDSA social worker about what strategies may help with cognitive and behavioral concerns. The following is a comment from a person who utilized a support group for help addressing problems.



#### *FAMILY NOTE*

*I sought help and guidance from other caregivers. As each problem arose, I tried to find ways to make life easier to reduce the difficulty.*

#### HDSA SUGGESTS:

- Talk to a social worker for support and resources.
- Talk to your person with HDs' primary doctor or HD specialist.
- Read information about the behavioral elements of the disease.
- Attend a support group.
- Watch a webinar on the topic of cognitive difficulties.
- If you are feeling unsafe, call 911 and ask if there is a Crisis Intervention Team (CIT) in your community that will help to address behavioral concerns.
- Attend a local or the HDSA national convention to learn more about HD, hear about novel treatments, listen to professionals talk about caring for a person with HD and to meet other HD families that share common concerns.



## Resources

Find a social worker in your area. Use the locator button [www.hdsa.org](http://www.hdsa.org)

*Understanding Behavior in Huntington's Disease: A Guide for Professionals*

<http://hdsa.org/wp-content/uploads/2015/03/Understanding-Behavior.pdf>

*A Physician's Guide to the Management of Huntington's Disease*

HDSA.org website. <http://hdsa.org/shop/publications/>

Advanced Directives

[http://hdsa.org/wp-content/uploads/2015/03/HDSA\\_Family-Guide-Advance-Directives-for-HD.pdf](http://hdsa.org/wp-content/uploads/2015/03/HDSA_Family-Guide-Advance-Directives-for-HD.pdf)

Caregiver Corner webinars

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/>

Palliative Care

We Are HDSA

[http://hdsa.org/wp-content/uploads/2015/03/1\\_We-Are-HDSA-Dec-2014.pdf](http://hdsa.org/wp-content/uploads/2015/03/1_We-Are-HDSA-Dec-2014.pdf)

Community resources

[Archrespice.org/respitelocator](http://Archrespice.org/respitelocator)

Aging and Disability can be found by searching for Area Agency on Aging (that supports individuals who are disabled as well as aging.) [www.n4a.org](http://www.n4a.org)

U.S. Department of Health and Human Services, Administration on Community Living <http://www.aoa.gov/> and also to find local resources <http://www.eldercare.gov/>

Disability

Caring Voice Coalition [www.caringvoice.org](http://www.caringvoice.org)

Legal

National Academy of Elder Law Attorneys <https://www.naela.org>

Long Term Care

<http://hdsa.org/wp-content/uploads/2015/05/long-term-care.pdf>

<https://www.medicare.gov/your-medicare-costs/help-paying-costs/pace/pace.html>

American Association for Long Term Care Insurance website might be helpful. <http://www.aaltci.org/>

[www.longtermcare.gov](http://www.longtermcare.gov)







# Chapter 4

## Occupational Therapy for a Person with Huntington's Disease



# Occupational Therapy for a Person with Huntington's Disease

*Are you a caregiver wondering about stages of HD and how well your person with HD can manage his finances, do the yard work or her ability to care for her children or how to assist her in the shower?*

*Please read this chapter about how an Occupational Therapist can assist and accommodate a person with HD to find comfort and safety in the home environment through all stages of HD.*

## The Role of the Occupational Therapist (OT) in the Healthcare Team

Occupational therapy, as defined by the American Occupational Therapy Association (AOTA), is “the therapeutic use of everyday life activities (occupations) with individuals or groups for the purpose of enhancing or enabling participation in roles, habits, and routines” within any environment of a person’s daily life (2014, p. S1). Everyday life activities include personal self-care tasks, home management tasks, and community activities. Accordingly, Occupational Therapists (OTs) see patients in many settings including outpatient clinics, nursing facilities, mental health programs/facilities, or through home health. Individuals who receive home health OT are homebound and may live in traditional homes, independent living facilities, or assisted living facilities.

As Huntington’s disease (HD) causes gradually impaired cognition, loss of physical control of movements, and behavioral issues, OTs are well-trained to provide guidance to maximize safety while promoting independence to ensure quality of life across the stages of HD. The OT’s job is to help a person with HD and their family and/or caregivers determine ways for the person with HD to live his/her life as fully as possible by understanding the person, his/her lifestyle, and typical disease progress. Multi-disciplinary clinics at a Huntington’s Disease Society of America Center of Excellence (HDSA COE) will include an OT as part of the outpatient healthcare team, who will see a person with HD in the clinic and/or act as an ongoing resource to the person with HD, the family and/or caregivers, or other healthcare providers. OTs are also available through home health care and other outpatient settings.

For more information on OT, please visit [www.aota.org](http://www.aota.org)

## The Guideline for Family and Caregivers of Persons with Huntington's Disease

*OT can help YOU help your person with HD to live life to the fullest!*

This guideline is intended to give general occupational therapy-based guidance for the families and caregivers of persons with Huntington’s Disease (HD). One of the most important parts of occupational therapy is for the occupational therapist (OT) to become familiar with the person with HD and his/her family and caregivers in order to provide personally meaningful advice. In the following, the pronoun “you” will be used to refer to the individuals, family members, and caregivers, who are the day-to-day support for a person with HD.

**This guideline should not replace working directly with an OT.** OTs who specialize in working with persons with HD are a part of the healthcare team at many HDSA Centers of Excellence (HDSA COE), but there are over 110,000 OTs in the United States (U.S. Bureau of Labor Statistics, 2014). The HDSA COE locations are shown on the HDSA website: <http://hdsa.org/about-hdsa/centers-of-excellence/>. However, some persons with HD may wish to see a local OT more frequently. For example, people who are receiving short-term rehabilitation in a facility may see an OT almost daily, but others may only need to check-in with their OT at the HDSA COE or outpatient clinic. It all depends on the needs of the person with HD. See the chapter on social services that discusses ways to pay for services.



Persons with HD can show declining abilities in thinking, coordinating their movements, and in maintaining calm behavior. Each person with HD can have varying degrees of difficulty and decline in each of these areas, and each person's symptoms progress at a different speed. Every person is unique, and the way each person with HD lives their life to the fullest is unique too. The starting point for any OT helping your person with HD is understanding how each of these areas (thinking, coordination, and mental health) effects your person with HD at his/her current life stage. This is particularly important for figuring out how daily life tasks can be safe, appropriate, and meaningful. For example, consider that a person with HD who is newly symptomatic could be in his late 20's, having just started a career, and have a young family; this person may be most concerned about safety and independence for driving, working, and caring for his children. Another person with HD who is newly symptomatic may be finishing his career, preparing for retirement, and planning to work in his yard and travel with his wife; he may be most concerned about his safety and independence in doing yard work and travelling. How the symptoms of your person with HD progress, and his/her life stage, will greatly influence how and whether the following suggestions apply.

The progression of HD symptoms follows a basic trend, and HDSA has provided the following brief descriptions of the three general stages of the HD.

**Early stage HD** usually includes subtle changes in coordination, perhaps some involuntary movements (chorea), difficulty thinking through problems and often a depressed or irritable mood. . . . The effects of the disease may make the person less able to work at their customary level and less functional in their regular activities at home.

In the **middle stage**, the movement disorder may become more of a problem. . . . [and] changes in thinking and reasoning abilities [occur]. . . . Ordinary activities will become harder to do.

In the **late stage**, the person with HD is totally dependent on others for their care. . . . Chorea may be severe or it may cease. At this stage, the person with HD can no longer walk and will be unable to speak. However, he or she is generally still able to comprehend language and retains an awareness of family and friends. (HDSA, Mid to Late Stage HD, 2015)

The following sections will refer to these three stages of HD in order to help you find strategies that are most helpful for your person with HD.

For nearly all persons with HD, there are a few core ideas that support the specific suggestions for independence and safety. First, and often foremost, the benefits of a regular routine cannot be understated! A consistent daily routine helps with memory, knowing what to expect, using safe strategies, and lessening anxiety and frustration. The consistency in the daily routine should include when, with whom, and how daily tasks are done.

*Family Note*

*Following a routine for showering and eating was important with my loved one.*

Secondly, modifications to keep the home environment quiet help to reduce distractions, stress, and anxiety. Thirdly, it is helpful for the home to be tidy and well-organized, as this can help with memory difficulties. Lastly, an accessible home reduces fall risk and increases independence. Ideally, a home will not have steps or stairs at the entrance or within the home, a large open bathroom, a walk-in shower, and grab bars installed in the shower and next to the toilet. In some home layouts, it is helpful to install handrails in hallways too.

*Family Note*

*We equipped our home with a high toilet and rails in the shower.*

*We had our bedroom and bath renovated to accommodate her disability; no stairs, many hand and wall railings.*



One of the most critical conversations among the person with HD, the family and/or caregivers, and healthcare professionals is determining the appropriate level of supervision for the person with HD. Most persons with HD need little or no supervision when they are initially symptomatic (early stage), while most persons with HD will need continuous supervision when they are in the late stages of HD. In general, as symptoms progress, the required level of supervision progresses too. Persons with HD have many variations in symptoms and rate of progression, so there are many variables to consider when deciding how much supervision a person with HD needs in order to safely and independently perform activities of daily living. The following are some questions to consider when thinking about the level of supervision your person with HD needs:

- Can he consistently and appropriately manage his bathroom needs? If there are problems with incontinence, can he safely clean himself as needed?
- Can he consistently recognize a safety problem, know how to appropriately respond, and be able to respond? For example, would he notice that cigarette ash fell on the carpeting and started a small fire? Would he be able to quickly figure out who to call for help? Can he find a phone and coordinate his movements to dial a phone?
- Will he remember and follow directions about things he should not do on his own (e.g. driving, operating machinery, cooking - as appropriate)?
- Will he attempt to leave the home or yard and not be able to safely manage in the neighborhood around vehicles? Can he remember where he is going and how to return home consistently?

If you, any of the family members, or caregivers believe that your person with HD cannot consistently do any one of these things then, it is time to discuss, as a family, and with the medical team, the ways to increase supervision. Persons with HD, their families/caregivers, and medical teams should regularly discuss the amount of supervision needed. The degree of need for supervision gradually increases with the progression of symptoms. For some persons with early-stage HD, it may mean that a family member or friend visits during the daytime to check that the person with HD eats lunch and takes his medication. Some persons with HD and their families/caregivers are able to extend supervision through the use of in-home video monitoring or even just a video baby monitor (Note: It is important to check the distance range on the video baby monitor; it may allow a caregiver to “keep an eye on things” from the other end of the house, such as while doing laundry, or while doing yard work). Other families have successfully used an alarm system in the house. Some persons with HD and their families/caregivers pursue adult day programs or community clubhouses; these programs can vary greatly in services provided and costs. Some persons with HD and their families/caregivers make plans for in-home care, either through private pay or Medicaid. Please remember that the OT is most helpful in the decision-making about when to increase supervision and how much supervision is needed, and the social worker can help to organize and plan for increased supervision through services and programs.

The following sections will emphasize general recommendations for improving safety and independence in daily occupations for a person with HD in early-, mid-, and late-stage. Occupations include activities of daily living (ADLs), which are self-care tasks, and more complex activities accomplished in the home and community. Please remember that the following guide is just a start and that working personally with an OT will help your person with HD and you maximize quality of life.





Occupation

# Working

|             |   |
|-------------|---|
| Stage of HD | <p><i>General Notes:</i> Since many see their work as a part of their identity, it is often a challenging decision for a person with HD to decide to adjust work duties or stop working completely. Decisions about work can be difficult because of concern about household finances. Please note that stress, such as stress from struggling to perform work duties, can make HD symptoms seem worse. Many persons with HD and their families/caregivers have noticed that the decision to stop working greatly reduced their stress and that the person with HD functions better. The HDSA COE social workers are very helpful in sharing resources that are available as well as how to apply for disability.</p>   |
| Early-Stage | <p>Consider whether the person with HD can <b>adjust his work duties to improve safety</b>. For example, if a short-order cook is getting burned from bumping the oven or stove while rushing, he would be safer doing prep cooking/baking rather than short-order style cooking. Or, it could be appropriate for this restaurant worker to start managing the salad bar to completely avoid possible burns. In other jobs, it may mean delegating certain higher risk tasks to another employee. These types of adjustments to work duties may be more realistic if the work supervisor has some level of understanding of the HD diagnosis.</p> <p>For both the person with HD and/or family members: Consider <b>adjusting work hours to reduce personal or household stress</b>. This may mean shifting to a more consistent routine for the family. It could also mean inquiring about the ability to do some work from home. Or, it may mean reducing overall work hours, such as having a half-day mid-week each week.</p> <p>Consider whether the person with HD can work with a supervisor to make <b>changes at work to reduce distractions</b>. For example, if the office is loud and busy, can the desk be moved to a corner space or a space where the person with HD can close the door when trying to focus? Perhaps, the office is particularly loud between 8-10 AM each day. Can the person with HD begin work at 10 AM to avoid these distractions?</p> <p>Consider whether the person with HD can do <b>this same job at a closer location to home to reduce stress</b> from the burden of a longer commute. For example, if the person with HD works in a grocery store, consider if that same grocery store has a location closer to home.</p> <p>Consider whether the person with HD can <b>change her job to reduce the stress</b>. For example, an elementary school teacher with early stage HD symptoms may be overwhelmed with trying to plan lessons and manage a large class of students. She could reduce her stress and daily challenges but still be able to earn an income by becoming a teacher's assistant rather than a head teacher.</p> |
| Mid-Stage   | <p>Many persons with mid-stage HD have either recently stopped working or are in the process of doing so. <b>The decision to continue to work requires that the person with HD be in a highly supportive and flexible work role without safety concerns for the person with HD, his co-workers, or any customers/clients. Keep in mind that safety means NO harm, not merely prevention of physical harm.</b> For example, an error on a financial statement would not cause physical harm to a client but it could cause significant harm to the client's business or personal finances. It is time to plan to stop working when the person with HD cannot safely and effectively perform work duties.</p> <p>Remember that <b>reduced stress, as from not working, can notably improve functional abilities.</b></p>  |
| Late-Stage  | <p>Working is not appropriate.</p>  |



## Driving/Community Transportation

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| Stage of HD | <p><i>General Notes:</i> Driving requires advance abilities that include the ability to pay attention, anticipate problems, remember in-the-moment where cars are located around your vehicle, and appropriate control of the steering wheel and pedal(s). The ability to drive represents independence to many, which is why many people are sensitive about their driving ability.</p>   |
| Early-Stage | <p>Consider <b>reducing distractions</b>, such as adjusting the radio when the vehicle is stopped and/or keeping the radio at a low volume..</p> <p>Drive <b>below the speed-limit</b>. Consider avoiding driving at high speeds, such as on the interstate.</p> <p>Avoid congested traffic. Run errands during off-peak traffic times.</p> <p>Use <b>cruise-control</b>, as appropriate, to help minimize pulsing of the gas pedal.</p> <p>Do not follow other vehicles closely. Always <b>leave extra space around, and especially in front of, the vehicle</b>. HD can impair some people's judgment of distance and force. It is important for a person with HD to have extra time to respond to any sudden changes in the traffic.</p> <p>A GPS can be helpful (helping the person with HD stay attentive to turns) or harmful (distracting the person with HD from focusing on the road); this depends on the person.</p> <p>Consider <b>removing distractions</b>, such as leaving the radio turned off, placing the cell phone in an out-of-reach location, and instructing passengers to remain quiet.</p> <p>Choose a family member or friend that the person with HD trusts to <b>regularly ride with and follow behind the vehicle while the person with HD is driving</b>. It is generally recommended that this happen monthly.</p> <p>Often, the decision to stop driving is gradual. The person with HD may limit where, when, how, and how much he drives before stopping altogether.</p> <p>Initiate a <b>family conversation</b> about how the person with HD would prefer to plan for transportation when driving is no longer an option. As the disease progresses, it can be more difficult for the person with HD to be able to exercise good judgment and/or effectively communicate his wishes about this topic.</p> <p>Handicap-accessible parking and/or parking close to an entrance can be helpful for some persons with HD at some stages. However, often when the early-stage person with HD is still driving independently, it is often less safe and more difficult to park close to an entrance. Parking close to the store entrance means parking and backing up in a more congested area (an area with more vehicles and more pedestrians). It can be harder to focus on controlling the vehicle in the tighter space, and there can be a higher likelihood of almost striking a pedestrian, especially if easily distracted. In general, it is recommended that the person with HD park in an open area of the parking lot with fewer vehicles and pedestrians and to pull-through when parking to avoid backing-up. Often, if a person with HD cannot safely walk to a store entrance, then it is likely that he does not have the physical ability to safely drive.</p> <p>If the person with HD normally relies on public transportation (subway, buses, etc.), it is still important for a family member or friend to regularly tag-along to see that the person with HD is continuing to use good judgment around traffic and appropriately managing public transportation schedules and/or changing of buses or trains.</p> |
| Mid-Stage   | <p>If family members or caregivers no longer feel safe riding with the person with HD or no longer feel that it is safe for the person with HD to drive children or grandchildren, it is time to consider how to proceed with helping the person with HD decide to stop driving. The three main ways to help the person with HD to stop driving are 1) the person with HD and family/caregivers decide together, 2) the person with HD goes to have a driving test at the Department of Motor Vehicles (DMV), or 3) the person with HD goes to have a driving test with a Certified Driver Rehabilitation Specialist (CDRS). It is important for the person with HD to be a part of this often sensitive decision.</p> <p>(<a href="http://myaota.aota.org/driver_search/index">http://myaota.aota.org/driver_search/index</a>) An OT maybe a CDRS who will emphasize both safety and independence in driving, including providing adjustments for driving (rather than simply a yes or no to being able to drive). A larger listing of those who are a CDRS can be found on this website: <a href="http://www.aded.net/">http://www.aded.net/</a>. Call ahead to learn more about the costs of the different programs available. Consider that having documentation indicating that the person with HD demonstrates safe driving habits could be helpful in the case of an accident.</p>  |
| Late-Stage  | <p>Driving is not appropriate.</p>   |



Occupation

# Child Care

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| Stage of HD | <p><i>General Notes:</i> Caring for children requires advanced thinking, physical, and emotional control. This applies whether the person with HD is involved in parenting or providing child care for family, friends, or customers. There are many safety concerns involved depending on the age of the child or children and the symptoms of the person with HD. A parent with HD is encouraged to remain involved in parenting tasks and spend time with his/her children even when he/she cannot be the primary responsible parent. However, measures must be taken to ensure that no harm comes to any child.</p> <p>HD symptoms can be confusing for children, and it can be difficult to know what to share when talking to children about HD. The HDSA National Youth Alliance provides support and resources for children affected by HD: <a href="http://www.hdsa.org/nya/">http://www.hdsa.org/nya/</a></p>  |
| Early-Stage | <p>If the person with HD has difficulty focusing on tasks and remembering, then a <b>second responsible adult should be present</b> while the person with HD is participating in the care of a child. This is critical for young children, such as babies, toddlers, and children in elementary school, and it remains important until the child is mature enough to be responsible for basic safety on his/her own.</p> <p>If the person with HD has mild difficulty with balance, then she should <b>avoid carrying the child in her arms</b> or in an on-body carrier while walking. Instead, consider using a stroller in the home to move the child from room to room.</p> <p>If the person with HD has mild difficulty with unintentional body movements (such as chorea) and wants to hold a young child, then encourage the person to <b>sit in a very supportive chair</b> (like a large recliner) and have the child placed in his lap.</p> <p>If the person with HD has mild difficulty with planning, problem solving, and/or remembering, often <b>another responsible parent or caregiver needs to be involved in scheduling the children's</b> plans for travel to/from school or extracurricular activities.</p> |
| Mid-Stage   | <p>Likely the person with mid-stage HD <b>will need supervision while participating in activities with children</b>, particularly if thinking and/or emotional abilities are becoming more limited.</p> <p>The person with HD may be able to participate in directly supervised <b>play with children sitting or lying on the floor</b>, as this position prevents the person with HD from falling and harming himself or the child during play. "Rough-housing" is not appropriate.</p> <p>A parent with mid-stage HD may still enjoy watching a child play sports or perform in a recital, and it may be appropriate for the <b>parent to watch the activity or be part of the activity</b>. Plan ahead for managing toileting concerns (urgency and/or incontinence), especially since not all public spaces have family-style bathrooms. An alternative could be for the parent with HD to attend a practice session or a dress-rehearsal.</p>   |
| Late-Stage  | <p>A person with late-stage HD may appreciate short visits from a child that he or she knows well. <b>It is important that the visit be a positive, safe experience for both the person with HD and the child</b>. Safety can be a concern if the person with HD has issues with controlling his/her emotions. The level of appropriateness for planning a child visit with someone with late-stage HD greatly depends on the age and maturity of the child and the types of symptoms that the person with late-stage HD exhibits. Caregivers should discuss these possible concerns before planning a visit.</p>  |



Occupation

# Household Finances

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| Stage of HD | <p><i>General Notes:</i> Household finances includes tasks like keeping bills organized, keeping track of household income and accounts, and budgeting for expenses appropriately. Good management of household finances requires attention, organization, memory, and problem solving.</p>   |
| Early-Stage | <p>If the person with HD has typically managed the household finances, it is a good idea for <b>him to teach a second responsible adult about the household bills and accounts.</b> This makes it easier for the second person to help if or when needed.</p>   |
|             | <p>If the person who has HD has typically managed the household finances, it is wise for <b>second person to be a “double-checker”</b> periodically to help avoid an error that might affect the household finances. It is valuable for the person with HD to stay involved in the planning and managing of the household as long as they can do so appropriately.</p>  |
|             | <p>A person with HD who has mild difficulty with attention, memory, and organization may consider getting his <b>primary bills scheduled with auto-pay</b> through a bank account.</p>  |
|             | <p>A person with HD who is still working and the family may find it simpler to use <b>auto-deposit of paychecks.</b> This keeps paychecks secure so that it cannot get lost.</p>  |
| Mid-Stage   | <p>A person with mid-stage HD symptoms regarding attention, organization, memory, problem solving, and judgment, will likely need <b>another responsible adult to manage the major household bills and accounts.</b></p>  |
|             | <p>It is appropriate and valuable for the person with HD to retain a <b>smaller personal budget for personal expenses,</b> such as going to the movies with a family member or being able to buy a snack when shopping with the family. Depending on the household finances, it may be easier to put some money on a prepaid debit card or to use cash. It is generally more difficult to manage money using a credit card or check book, especially for someone who has any difficulty with attention, memory, or problem solving.</p> |
|             | <p>It is valuable to <b>communicate household financial decisions with the person with HD.</b> It is particularly challenging for many persons with HD to adjust to change, especially if the change seem sudden. For example, if the family needs to move to a new home because of family income, it is important that everyone knows that this move was thoughtfully considered and that this is communicated well ahead of the move.</p>   |
| Late-Stage  | <p>Household finances should be managed by another responsible adult.</p>   |





| Occupation         |  |
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| <h1>Yard Work</h1> |  |
| Stage of HD        | <i>General Notes:</i> Yard work includes many different activities of varying difficulty, such as mowing, weeding, raking, shoveling snow, and tending to a garden (planting, watering flowers, and weeding). In general, doing yard work requires good balance, strength, and energy. However, it also involves good judgment, particularly when using any motorized lawn-care equipment or tools. Some yards are hilly and uneven, which are more challenging for a person with decreased balance.   |
| Early-Stage        | The <b>use of motorized lawn-care equipment or tools</b> should be generally supervised (semi-regular supervision at a minimum) to ensure that good judgment and consistent attention to safety is being used by the person with mild HD symptoms. While mowing with a riding lawnmower is usually safer than driving a car, distraction can be a safety concern that can result in rolling a lawnmower on a hillside or spilling gas.   |
|                    | The <b>use of ladders is strongly discouraged</b> due to risk of injury from falling whether the mild HD symptoms are related to difficulties thinking and/or difficulties with balance. This applies to cleaning gutters, hanging holiday decorations, or pruning trees.  |
|                    | Safety while doing yard work <i>greatly</i> depends on the extent of the symptoms of the person with HD. <ul style="list-style-type: none"> <li>• A person with mild balance issues should avoid tasks that challenge balance, such as shoveling snow and blowing leaves. Instead, he could do less physically demanding tasks, such as watering plants, weeding, or filling a bird feeder.</li> <li>• A person with mild thinking impairment but not balance issues could be able to safely accomplish tasks like raking leaves or shoveling snow.</li> </ul> |
| Mid-Stage          | The use of <b>motorized lawn-care equipment or tools is inappropriate</b> for nearly all persons with mid-stage HD symptoms.   |
|                    | A person with mid-stage HD <b>may be able to safely accomplish simple, light yard work tasks</b> , such as tending to a container garden or filling a bird feeder. Supervision may be needed to help the person with HD remain on task, but also a person with mid-stage HD symptoms may not be interested in yard work.   |
| Late-Stage         | Yard work should be managed by another responsible adult.  |



| Occupation        |   |
|-------------------|---|
| <h1>Cleaning</h1> |   |
| Stage of HD       | <i>General Notes:</i> As cleaning involves many tasks of different difficulty levels, safe and effective performance depends greatly on the symptoms of the person with HD. Considering everything from vacuuming, washing floors, and scrubbing the bathroom to doing laundry and washing dishes, it is important to think about which tasks require good balance and/or good attention and memory.  |
| Early-Stage       | In order to <b>remember to do certain housecleaning tasks regularly and to divide them up throughout the week</b> , consider assigning certain tasks to certain days of the week. It can be a helpful reminder to use the memory strategies of alliteration or rhyming. For example, Mondays - "Must dust," Tuesdays - Toilets, Wednesdays – Wash clothes, Thursdays – Tubs, and so on.   |
| Early-Stage       | For anyone with balance problems, <b>always have one hand on a handrail when going up and down stairs</b> , especially when carrying something. Persons with HD with even slightly decreased balance, are discouraged from carrying large items with both hands up and down any stairs because of the increased risk of falling/injury. Anyone with balance problems should consider using a small laundry basket that can be carried under one arm or use a laundry bag with a shoulder strap if laundry has to be carried up and down stairs. Similarly, a person with balance problems should avoid carrying a bulky, heavy vacuum up and down stairs. Instead, they could use a lightweight vacuum that could be carried in one hand (think of the dust buster on a long handle style) or they could ask someone else to carry it up or down stairs for them. |
|                   | For anyone with any balance problems, and especially anyone who also has problems with short term memory, <b>use extra caution when vacuuming so that the power cord does not become a trip hazard or a wet floor become a slipping hazard.</b>   |
|                   | Some persons with HD have difficulty completing tasks because of problems with attention and memory. These people may leave wet laundry in the washer until the clothes start to smell. One strategy may be to <b>set a timer with an alarm on a phone, a watch, or digital kitchen timer</b> to alert the person to return to complete the task. For example, keep a digital kitchen timer on top of the washer and always start it when a load of laundry is started, and then return to the washer when the alarm goes off to remove the wet clothes from the washer.  |
|                   | Some persons with HD have difficulty starting tasks because of problems with initiation. They want to get something done and know how to do it but they cannot seem to figure out how to get started. This can be overwhelming and frustrating for the person with HD and his/her family. They may do better if they have a <b>very consistent routine of household chores, or just simple tasks</b> , or they may need gentle, supportive reminders to start the task. It is valuable to remember that this is a common symptom of HD and not just laziness.   |
|                   | Some persons with HD have difficulty noticing what tasks need to be done or organizing their day to do those tasks because of problems with thinking (executive processing). <b>It can be helpful for the caregiver to leave the person with HD a short, reasonable list of "to-do's" in the same obvious, central place every day.</b> Remember that the list must be concise and easily accomplished.   |
| Mid-Stage         | A person with mid-stage HD symptoms may be able to safely accomplish simple, light household cleaning tasks, such as putting unbreakable dishes into the dishwasher, wiping the counters, or folding laundry (depending on level of chorea). Supervision may be needed to help the person with HD remain on task, but also a person with mid-stage HD may not be easily engaged in household cleaning tasks. It is important to only encourage tasks that are safe and easily accomplished to keep the person with HD feeling successful.   |
| Late-Stage        | Household cleaning should be managed by another responsible adult.  |



Occupation

# Cooking

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| Stage of HD | <p><i>General Notes:</i> Cooking full meals using the stove, oven, or grill requires good safety awareness, attention, problem solving, short term memory, coordination, and balance. Making cold meals, like sandwiches, or heating food in the microwave requires all of the above abilities but to a lesser degree; so these types of food preparation are easier and often safer. The key points for safety are appropriately managing use of heat and the safe use of knives.</p>   |
| Early-Stage | <p>Some persons with HD who have difficulty remembering to complete tasks may leave the oven or grill on after removing food. Anytime heat is involved, forgetting to turn it off can be a concerning safety issue. One strategy can be to <b>set a timer with an alarm</b> on a phone, a watch, or digital kitchen timer to alert the person to return to the task. It's important that the timer/alarm can only be turned off manually. If the timer idea does not work for tasks that involve heat, then it is good to have another person double-checking that the heat source is turned off for safety.</p> <p>Another option for persons with HD who have difficulty remembering to turn off the oven is to use a <b>counter-top toaster oven that has a built-in timer with an automatic shut-off function.</b></p> <p>In order to make it easier to remember where to find items in the kitchen and where to put them away, consider placing <b>labels on the cabinets and drawers in the kitchen.</b></p> <p>Some persons with HD who have difficulty with short-term memory have difficulty remembering which ingredients they have already added in a recipe. It can be helpful to <b>create a routine for organizing the ingredients</b> – some will put away the ingredient after they have used it, some people will move the ingredient to the other side of the counter, and other people will photocopy the recipe and strike out the ingredient after putting it in. Stick with the strategy that works best.</p> <p>Some persons with HD who have difficulty with coordination cannot safely use a knife. Another option can be using <b>kitchen scissors</b> for cutting up thin meats (like thinly sliced chicken for a stir fry).</p> <p>Some persons with early-stage HD have very mild coordination issues and can be at a higher risk for bumping the inside of the oven when placing or removing items. Try using <b>extra-long oven mitts</b> (which cover most of the arm rather than just the hand and wrist).</p> <p>For persons who have difficulty with organizing and meal planning, it <b>can be helpful to have a regular weekly meal plan.</b> For example, spaghetti on Mondays, tacos on Tuesdays, stir-fry on Wednesdays, hamburgers on Thursdays, and so on.</p> <p><i>Related Note:</i> Grocery Shopping – Some people may find grocery shopping overwhelming because of difficulty with memory and/or decision-making. In this case, it is helpful to <b>always use a grocery list with the items listed specifically</b> (so “4 pears” rather than writing “fruit”) or writing the preferred brand (which can help prevent worrying over which brand to get). For an additional charge, some stores offer online shopping options that can be home delivered or picked up at the store.</p> |
| Mid-Stage   | <p>Many persons with mid-stage HD have difficulty cooking meals and safely using the stove, oven, or grill. These individuals can more safely use the microwave to heat foods. For some who have with difficulty with attention, problem solving, and decision-making, it can be challenging to use the <b>microwave</b> because of the number of buttons. If this is a concern, <b>a mark or a sticker placed on the “Add 30 seconds” button</b> can make it very visible and then leftovers can be marked with the number of times to push the “Add 30 seconds” button. For example, a star can be marked on the “Add 30 seconds” button and then the leftover pizza could have “2 *s” written on masking tape on the Tupperware holding the pizza. Sometimes the “Start” button needs to be marked too, depending on the type of microwave.</p> <p>Some persons with HD and their families/caregivers find it easier for there to be a <b>fully-organized cold lunch</b> (even with napkin and utensils) placed in the same place in the refrigerator for meals when there are alone.</p> <p>For persons with HD who cannot safely use the stove or oven and do not recognize this, it may be necessary to <b>remove the stove knobs or unplug the appliance</b> when the person does not have direct supervision.</p>  |
| Late-Stage  | <p>Cooking should be managed by another responsible adult.</p>   |



| Occupation                   |   |
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| <h1>Managing Medication</h1> |   |
| Stage of HD                  | <i>General Notes:</i> Medication compliance is critical to managing symptoms of HD. This requires problem solving, planning, memory, and fine motor coordination, as well as good vision.   |
| Early-Stage                  | Nearly everyone with HD who has been prescribed medications should get in the routine of <b>writing on the bottle or a calendar when a prescription should be finished</b> , and then check on that date if there are any pills left. This is a good way to double-check that doses have not been skipped.  |
|                              | Some persons with HD find it helpful to place their prescriptions on <b>automatic refill</b> at the pharmacy, to reduce the chance of a lapse in prescriptions.   |
|                              | Remembering to take medications is easier if it fits into a routine. For instance, people who make coffee every morning may put their morning medications or their pill box next to their coffee pot to help them remember their medications.   |
|                              | Persons with HD who have mild difficulty with memory and organization often find it helpful to organize their pills in a <b>pillbox</b> for each week. Some pill boxes have just one space for pills for each day but others have a slot for AM and PM or slots for breakfast, lunch, and dinner. It is good to look for a pillbox that fits the schedule of the prescriptions. If all pills are taken at the same time, then a pillbox with just one space for pills per day is appropriate. |
|                              | For persons who have difficulty with attention and memory, it can be helpful to use an <b>alarm</b> as a medication reminder. This can be done by putting an alarm clock next to the pill box, setting an alarm on a personal cell phone, or purchasing a pill box with a built-in alarm.   |
|                              | To avoid dropping pills, it is helpful for some people with mild coordination problems to <b>sit down and prop their forearms or elbows on a table</b> while taking or sorting pills.   |
| Mid-Stage                    | Some persons with HD may be able to swallow pills better when the <b>pills are placed in food</b> , such as applesauce or oatmeal. When considering this last option, it is helpful to check with your medical team, especially the speech-language pathologist to check on any food consistency recommendations.   |
|                              | Persons with mid-stage HD often require <b>someone else to remind them</b> to take their pills. For some people, a phone call and reminder is enough for them to consistently take their pills but others may require a person to give them their pills.  |
|                              | To avoid dropping pills, it is helpful for some people with moderate coordination problems to <b>sit down</b> while taking pills that have been placed in a <b>firm, unbreakable cup</b> . The person with HD can use the cup to bring the pills to the mouth (like drinking from a cup).   |
| Late-Stage                   | Medications should be managed by another responsible adult.   |



Occupation

# Showering

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| Stage of HD | <p><i>General Notes:</i> Safe and effective showering requires balance, attention, and short term memory. For persons with HD who have decreased balance, it is important to alter the bathroom area to reduce the chance of falling. Persons with HD who have decreased attention and memory may have difficulty remembering to take a shower, getting started taking a shower, and following the sequence of washing (e.g. remembering if all areas of the body have been washed and rinsed). For these types of difficulties, compensations are shared below.</p>   |
| Early-Stage | <p>Some persons with HD have difficulty remembering and starting tasks; they may forget or skip bathing/showering regularly. It can be helpful to establish a <b>consistent daily routine</b> to help make this task a regular, expected part of every day. For example, it may work well for the person with HD to always bathe/shower immediately following dinner every day. This will help establish a routine that allows the person with HD to remember to bathe or shower.</p> <p>Some persons with HD have difficulty with attention and short term memory and can forget which areas have been scrubbed while bathing/showering and may get out before they are fully clean. It can be helpful to establish the <b>habit of always scrubbing in the same order</b>. For example, always “start at the top and work down;” so wash hair, then face, then neck, then chest, and downward.</p> <p>Nearly everyone with HD should get in the habit of <b>holding a sturdy surface or grab bar while stepping in or out of the shower or tub</b>. The earlier these become a regular habit the better in order to reduce falls in the future. In most shower areas, it is best to install at least 2 grab bars:</p> <ul style="list-style-type: none"> <li>• Place one vertical grab bar at the entrance to the shower (placed on the same wall as the shower head) to use when getting in/out of the shower.</li> <li>• Place a second grab bar diagonally on the interior wall. The diagonal bar should have the higher side closer to the shower head and the lower side toward the back of the shower; this position allows a person to use the higher side while standing and the lower side while sitting on a shower seat.</li> </ul> <p>Typical grab bars that are installed securely into the studs of the walls of the bathroom are the best for durability. It is possible to buy high-quality, reliable suction-cup grab bars but these can be expensive. The suction-cup grab bars will not adhere to tiled walls if the tiles are smaller than the head of the suction cups.</p> <p>For people who drop their soap in the shower, a <b>wash mitt</b>, a typical mesh sponge with a strap (using the strap around the wrist), or soap on a rope can be useful.</p> <p>When balance (and fall risk) is an issue, the person with HD should shower when someone else is present at home.</p> <p>Apply <b>no-slip adhesives</b> to the tub or shower floor so it is not as slippery when wet.</p>          |
| Mid-Stage   | <p>At times, it can be difficult for some persons with HD to appropriately control the water faucet and/or to notice a safety concern, such as the tub or shower water is too hot. <b>Adjust the water heater temperature</b> setting on the water heater to avoid the risk of a scald-type injury.</p> <p>For those who cannot remember how to thoroughly scrub, it is sometimes helpful to have an easy-to-read list posted in the shower area. However, more often the person with HD may <b>need a family member or caregiver present to give reminders</b> (like, “Wash your hair, please”).</p> <p>If the person with HD is unsteady when holding on to a grab bar to get in/out of the shower, <b>a physically-capable family member or caregiver may need to be available to assist</b>. Many people with balance problems find it steadier and easier to <b>sidestep</b> over the side of a tub while holding on the grab bar; this also allows space for the caregiver to stand closely too.</p> <p>If the person with HD is unsteady even when holding on to a grab bar while in the shower, it is often best that he/she use a <b>shower seat</b> for better safety and independence. Removable shower seats are usually either a shower chair (usually best in shower stalls) or a tub transfer bench (usually best in tub/shower combination). The tub transfer bench is designed to straddle the side of the tub; 2 legs go on the outside of the tub. The hard plastic shower seats seem to be more durable for people with notable chorea or a tendency to flop down when sitting.</p> <p>When using a shower seat, it is helpful to install a <b>height-adjustable showerhead</b> (good for aiming the spray while the person with HD does his own bathing) or a <b>handheld showerhead</b> (good for the family member or caregiver to be able to aim the spray while helping with bathing). It is often difficult for persons with HD who have coordination problems to direct a handheld showerhead themselves. Some handheld shower heads have a small spray on/off toggle switch on the shower head which can be helpful.</p> <p>Some persons with mid or late-stage HD have a lot of coordination problems, balance problems, and/or chorea, and they may benefit from wearing a helmet for head protection. Given the higher risk of slipping and falling in the shower area, it can be appropriate for the person with HD to wear a <b>vinyl helmet</b> while showering as it can get wet.</p> |





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| Late-Stage | Even when a person cannot physically do his/her own bathing, using a <b>consistent daily routine</b> and the <b>habit of always bathing in the same order</b> can promote cooperation and help minimize the person's refusals due to anxiety or stress.   |
|            | Change is disruptive! If a person with HD is living in a facility, facility staff members are encouraged to try to have as much consistency in routine of care and caregivers as possible for showering/bathing.  |
|            | Family members and caregivers of persons with late-stage HD should be cautious and use <b>good body mechanics for caregiver safety</b> when assisting with showering or bathing. It can be easy to strain or slip while reaching and helping in the shower.   |
|            | Some persons with late-stage HD need help with all aspects of bathing. It is important to determine if it is safer to do <b>sponge baths or bed baths</b> . In facilities and in homes with special roll-in showers, it may be possible to use a <b>supportive roll-in shower chair</b> . Some rolling shower chairs also provide tilt and/or reclining positions. For example, BRODA's® shower commode chair on wheels offers tilt and recline positioning, as well as a padded toilet seat. |

| Occupation        |   |
|-------------------|---|
| <h2>Dressing</h2> |   |
| Stage of HD       | <i>General Notes:</i> Getting dressed and undressed often requires balance, hand coordination, attention, and problem solving. Some problems getting dressed may include putting clothes on inside-out or backwards, or difficulty managing buttons, zippers, or snaps. Some people have difficulty keeping their balance while getting dressed. Throughout the stages of HD, continue to encourage the person to participate as much as they can, even if only to lift an arm.   |
| Early-Stage       | People who have difficulty with attention and problem solving may <b>need to practice looking for tags</b> before putting their clothes on in order to be more focused on which part goes "in the back." Sometimes, it can help for the person to get in the <b>habit of always checking themselves in the mirror</b> after getting dressed. At times, a family member or caregiver will need to give <b>reminders</b> .  |
|                   | Some persons with HD may share that they get overwhelmed by the choices of what to wear. It can be helpful to simplify their daily clothing choices <b>by hanging matching shirt and pants together</b> in the front of the closet.   |
|                   | If it is difficult to manage fasteners (buttons, zippers, snaps, or shoelaces), it can be helpful to sit <b>down and prop the arms on something</b> while working on the fasteners. For example, the person with coordination issues can prop his elbows and arms against his legs to stabilize his arms while tying his shoes. Another example is when a woman has difficulty putting on her bra, she can fasten it with the hooks positioned over her chest and with her arms propped against her sides; then she can rotate the back of the bra to her back before putting her arms in the straps.   |
|                   | If it is difficult to manage fasteners (buttons, zippers, snaps, or shoelaces), encourage the following <b>alternatives to fasteners</b> : <ul style="list-style-type: none"> <li>• Put a key ring or paperclip on zipper pulls to make them larger and easier to grasp.</li> <li>• Keep most of the buttons on a button-down shirt buttoned, even in the laundry, and just button/unbutton the top couple of buttons to put the shirt on like a pull-over shirt.</li> <li>• Use elastic shoelaces to make supportive sneakers into slip-on/off shoes.</li> <li>• When buying new clothes, try to get clothes with elastic rather than fasteners. This may include getting sports bras rather than traditional bras.</li> </ul> |
| Mid-Stage         | Most persons with mid-stage HD should do <b>nearly all their dressing tasks while sitting down</b> .  |
|                   | Most persons with mid-stage HD need <b>some help</b> getting dressed.   |
|                   | Many persons with mid-stage HD will <b>only wear easy-to-put-on clothes</b> , such as clothes with no fasteners, or they will need help with certain dressing tasks.  |
| Late-Stage        | When it becomes difficult for a person with HD to stand while someone is helping him get dressed, it is <b>often safest to put underwear and pants on while the person with HD is lying down</b> in the bed or a chair that fully reclines.   |





| Occupation         |  |
|--------------------|--|
| <h1>Toileting</h1> |  |
| Stage of HD        | <p><i>General Notes:</i> Safety and effective self-care are both important parts of a person’s ability to manage his toileting needs. Transferring between standing and sitting for toileting is often more challenging because the height of a toilet is often lower than typical chairs. Because the seat is low and there is frequently urgency to use the toilet, there is a higher risk of falling while toileting. Additionally, the risk of injury is often higher because of the hard surfaces in the bathroom, including the toilet, countertop, side of a tub, or a tile floor. Effective self-care for toileting is also important for hygiene. Many people with difficulty managing their toileting needs can be modest or embarrassed to let others know that they need help.</p>   |
| Early-Stage        | <p>For persons with HD who have difficulty with attention (focusing and noticing) it can be more difficult to recognize the urge to use the bathroom until it is nearly too late. This can lead to the person rushing unsteadily to the bathroom and having a higher likelihood of falling or result in bowel or bladder incontinence (“accidents”). For both of these reasons, it is important for the person with HD to get in a <b>daily routine of bathroom breaks</b>. This can include going to the bathroom when waking up, before every meal, and before bedtime even if the person does not feel the urge to go.</p> <p>For some persons with HD, toileting hygiene can be a challenge. They may need to be encouraged or reminded to wipe thoroughly. Sometimes the use of damp, flushable <b>toilet wipes</b> can be helpful.</p> <p>Men with HD who have decreased balance and/or coordination often have difficulty appropriately aiming their urine stream into the toilet. Urine spills can be a safety hazard with regard to falls. To reduce fall risk and to improve home cleanliness, <b>encourage men to sit while urinating</b>.</p> <p>If a person with HD has slight unsteadiness when sitting down on the toilet, it can be helpful to develop the habit of <b>consistently reaching back</b> with one hand to the toilet seat for steadiness.</p> <p>If the plastic bolts on the toilet seat frequently loosen, replace them with metal ones.</p>   |
| Early-Stage        | <p>For those with mild to moderate unsteadiness it can be helpful to install <b>toilet grab bars</b> and to raise the <b>height of the toilet seat</b>. There are different methods to make these helpful changes in a typical bathroom:</p> <ul style="list-style-type: none"> <li>• Install traditional grab bars on a nearby wall (placed closely to the sides of the toilet)</li> <li>• Install a toilet safety frame (can have grab bars only or grab bars on an attached raised toilet seat)</li> <li>• Place a 3-in-1 bedside commode over the toilet (without the bucket; use a splash guard to ensure all waste is directed only into the toilet bowl)</li> <li>• Install a “comfort height” toilet</li> </ul> <p>It can be helpful to have portable <b>urinals</b> available at night and for travelling to reduce issues with fall risk and urgency. During any travel, even to and from doctors’ appointments, a urinal can be an easy alternative to managing in public restrooms. Urinals are available for both men and women.</p> <p>Some women with HD may have difficulty managing menstrual products and personal hygiene because of coordination and/or attention difficulties. If a woman with HD is having difficulty physically managing a tampon or remembering to change this product appropriately, it may be necessary to transition to only using pads. If a woman with HD has substantial difficulty managing her personal hygiene, consider discussing with the physician pharmacological options to reduce this burden.</p> |
| Mid-Stage          | <p>Most persons with HD eventually require the use of incontinence <b>briefs</b>. It may be more easily accepted by the person with HD if briefs are purchased when incontinence issues are still only occasional and store them in the bathroom for use whenever she/he is ready to use them. There are many brands and styles of briefs that fit differently, so buy a few different types to find the preferred brand and style. When briefs are used, it can become more difficult to notice that the brief is damp and this can cause skin breakdown. It is always important for the skin to stay clean and dry.</p> <p>As incontinence issues increase, it can become even more important to have a consistent daily <b>toileting schedule</b>. A toileting schedule could be as frequent as every 2 hours during the day. Toileting schedules can be helpful in both the home and in a care facility for encouraging safety and good hygiene.</p> <p>Some persons with mid-stage HD have significant difficulty with controlling how quickly or firmly they sit down on the toilet, and sometimes this can cause damage to the toilet. Some families have found it helpful to use a <b>3-in-1 bedside commode over the toilet as it is cheaper to replace</b> that than the entire toilet.</p>  |
| Late-Stage         | <p>For people with significant chorea who rely on briefs, it may be safer to avoid trying to stand the person while managing soiled briefs over the hips. Instead, <b>have the person with HD lie down</b> (either in a bed or using a reclining chair, such as a BRODA® chair, flattened). Roll the person with HD from one side to the other while managing pants or briefs up or down. Briefs with side tabs reduce the caregiver burden of having to remove shoes and pants to change soiled briefs. However, if the person with HD prefers the briefs without side tabs, then fully remove the shoes and pants first. Next, slide new, clean briefs up legs while rolling the person with HD from side-to-side to pull briefs upward.</p> <p>Carefully tear or cut with scissors the sides of the soiled briefs to remove (rather than dragging the soiled brief down the legs). Attend to hygiene and promptly place new brief up over hips; continue to assist with rolling to reduce friction while sliding brief and pants up.</p>  |





| Occupation              |   |
|-------------------------|---|
| <b>Hygiene/Grooming</b> |   |
| Stage of HD             | <p><i>General Notes:</i> Grooming tasks include oral care, hair care, shaving, applying deodorant, and nail care. When a person with HD has difficulty with attention and memory, it can become more difficult to remember to do and how to do these tasks. A person with HD who has difficulty with coordination can have difficulty with effectively managing the items used for these tasks, for example holding the toothbrush and brushing her teeth.</p>  |
| Early-Stage             | <p><b>Establishing a consistent daily grooming routine</b> can be helpful. For example, the routine for the person with HD may be to always brush his teeth, then wash his face, and then comb his hair immediately after eating breakfast regardless of whether he is planning to leave the house that day. For some people, it can be helpful to tape a written list of the routine in an easily seen place as a visual reminder of the tasks to be completed.</p> <p>Occasionally, some persons with HD can be obsessive and compulsive about performing certain grooming tasks, such as brushing their teeth so hard that the gums bleed. This should be shared with the medical team so it can be addressed.</p> <p>For someone with HD who has difficulty coordinating her hand movements, it can be helpful to <b>lean against a nearby wall</b> and/or <b>prop an elbow</b> on the countertop while doing grooming tasks at the sink. For many people with hand coordination difficulties, it is easier to control hand movements when the upper arm or elbow is propped on or against a wall or countertop.</p> <p>Using an <b>electric toothbrush</b> can compensate for some difficulty with hand/arm coordination rather than using a regular toothbrush. The thickness and heaviness of an electric toothbrush can make it easier to hold or maneuver.</p> <p>An <b>electric shaver</b> can be easier to manage than a regular razor for those with hand coordination difficulties because the electric shaver is thicker and heavier. It is also less likely to cause nicks.</p> <p>For styling hair, it can be easier and safer for a woman with coordination difficulties to use a <b>hot air brush</b> (hair blow dryer shaped like a round brush).</p> <p>Try to <b>store all frequently used items in an easy-to-reach location</b> rather than having to reach, open, and pull to get things out every day.</p> |
| Mid-Stage               | <p>Some persons with HD who have limited focus and memory will need a family member or caregiver to <b>remind them</b> to do their grooming tasks every day.</p> <p>If someone's standing balance is poor, it is better to <b>sit down at the sink</b> while performing grooming tasks. In some people's home bathrooms, a person can sit on the toilet lid and reach the sink but in other bathrooms there is enough room to have a chair. Other people may decide to do their grooming tasks at the kitchen sink because there is more space.</p> <p>If a person's coordination makes it hard to put the toothpaste on the toothbrush, she can squirt the toothpaste directly into her mouth (as long as she does not have tendency to squirt too much). If a person with HD has difficulty spitting, she can <b>dip the toothbrush in mouthwash</b>, rather than toothpaste, to reduce the need to spit.</p> <p>Many men with HD who have difficulty with coordination need <b>someone else to shave them</b>.</p>   |
| Late-Stage              | <p>Generally in late-stage HD, the person will <b>need direction and physical help</b> to do nearly all of his grooming tasks. At this time, it is often easier to do these tasks with the person sitting in a very supportive chair and to bring the items to him.</p> <p>The caregivers for a woman with late-stage HD will often decide that it is easier to manage <b>shorter hair styles</b>. On occasion for someone whose chorea is mild, it can be helpful to use <b>a hair washing tray or funnel at the sink</b>.</p>   |



Occupation

# Eating

|             |   |
|-------------|---|
| Stage of HD | <p><i>General Notes:</i> People who have issues with coordination often have difficulty holding cups and utensils, as well as difficulty scooping/spearing food and bringing it to their mouths. These difficulties can cause frequent spills.</p> <p>*If there is any difficulty with chewing and swallowing food, which may include someone coughing during eating, it is very important to be assessed by a <b>speech-language pathologist (also known as a speech therapist)</b>. The speech therapist can give recommendations on appropriate food textures. Occupational therapists and speech therapists often work together to address concerns about eating.</p>   |
| Early-Stage | <p>A person with HD may be able to better focus on controlling his utensils in a less distracting environment. Turn off the TV or radio during meals to <b>reduce distractions</b>. This can also lead to less spilling.</p> <p>For those who have difficulty with coordination, it is better to <b>eat at a table</b> rather than in a recliner or on the couch. Generally, the best position is sitting in a sturdy chair scooted up against a sturdy table. It is also good for the person with HD to <b>prop her elbows or forearms on the table</b> to reduce extra arm movements.</p> <p>Some people with coordination and attention difficulties may often drop things in the kitchen. It is best to use durable, hard plastic cups, bowls, and plates and to <b>avoid using breakable dishes and cups</b>. Broken glass is a safety hazard.</p> <p><b>Certain types of utensils</b> can make it easier and less messy to eat and drink when someone has issues with coordination. Some helpful options are:</p> <ul style="list-style-type: none"> <li>• Use larger spoons or a soup spoon (makes scooping easier).</li> <li>• Bend the “neck” of the utensil so the top of the utensil is easier to aim toward the mouth.</li> <li>• Use spoons with thicker handles.</li> <li>• Try putting foam tubing around the utensil handle to make it thicker.</li> <li>• Use weighted utensils (gives the hand more weight to feel where and how the utensil is positioned in the hand).</li> <li>• Try firm, unbreakable plastic utensils if there is difficulty slowing the hand movement when putting the utensil in the mouth (as occasionally people will break or damage their teeth by hitting the utensil against their teeth during a sudden, unintentional hand/arm movement).</li> </ul> <p>Many persons with coordination issues have difficulty holding, picking up, and putting down a cup without spilling the liquid. <b>Plastic cups with lids, handles, and a fairly narrow spout</b> for drinking are usually preferred. Lids reduce spills from sloshing liquids. Handles reduce the chances of cups being dropped, especially if the person with HD can slide his/her fingers inside the handle and around the cup (so even if the fingers open unintentionally, the cup handle will stay around the palm of the hand, and likely keep the cup from falling). A fairly narrow spout helps to keep the liquid from pouring out too quickly.</p> <p>For those who push food off their plates, it can be better to use bowls, plate guards, and/or pie pans – something unbreakable with a <b>lip around the dish’s edge</b>. For persons who spill food because their plate slides around on the table, it can be useful to put <b>Dycem®</b> or <b>non-slip shelf liner</b> between the bowl/plate and the table.</p> |
| Mid-Stage   | <p>Some persons with HD have difficulty focusing and remembering and may <b>need reminders that it is time to eat a meal</b>, as they can find it easy to snack nearly all the time on the same few favorite foods. It is important to have enough calories eaten to make up for the extra calories burned by chorea.</p> <p>There are also <b>adaptive utensils, dishes, and cups</b> that can be used at different stages and with different types of food. Some additional options for more significant coordination difficulties are:</p> <ul style="list-style-type: none"> <li>• A weighted cup</li> <li>• A self-leveling spoon</li> <li>• Utensils that are weighted and have extra-thick handles</li> <li>• A universal cuff (a strap that fastens around the palm to hold the utensil)</li> </ul> <p>Some persons with notable chorea will keep unintentionally sliding their chairs back, away from the table while eating. One option to address this is to put the back of the <b>chair against a wall</b> and then slide the table up to him.</p> <p>If severe chorea is causing bruising when sitting at the table, consider <b>attaching foam or rubber on the edges of the table</b>.</p> <p>Restaurants and nursing facilities can have very distracting, busy dining rooms. <b>One way to reduce spills</b> during meal times in these types of spaces is to turn the person with HD to face the fewest distractions possible.</p>   |
| Late-Stage  | <p>Many persons in late-stage HD will <b>need assistance</b> to scoop food and bring it to their mouths for eating. Sometimes, they are still able to manage certain “<b>finger foods</b>” on their own. Assistance for eating may mean that the family member/caregiver places her hand over the hand of the person holding the utensil, or it may mean that the family member/caregiver manages the task.</p>   |



| Occupation     |  |
|----------------|--|
| <h1>Sleep</h1> |  |
| Stage of HD    | <i>General Notes:</i> A person's ability to sleep can greatly impact on his/her ability to perform all other tasks and activities safely and successfully. Persons with HD often have difficulty staying on a consistent routine of sleep and wakefulness. Also, HD medications may impact sleep patterns.   |
| Early-Stage    | <p>Many persons with HD have a tendency to stay up very late at night and sleep late in the morning. However, a person with HD who is a "night owl" is less likely to have readily available supervision and assistance. Consistent sleep routines are very helpful for everyone in the family. <b>Encourage the person with HD to wake at nearly the same time daily and to avoid shifting the sleep/wake schedule day-to-day.</b></p> <p>If the person with HD needs to call for assistance during the night, encourage them to <b>keep a cell phone near the bed.</b></p>   |
| Mid-Stage      | <p>Some persons with HD have a lot of activity during sleep and some have issues with rolling out of bed. Various soft methods can be tried to reduce the person's risk of falling out of bed:</p> <ul style="list-style-type: none"> <li>• Tucking a rolled blanket, body pillow, or a swimming pool noodle under the fitted sheet near the edge of the bed to make the middle of the mattress lower than the edges.</li> <li>• Considering buying a concave mattress.</li> </ul> <p>Please note that if rigid bedrails are necessary, they must be covered with soft material, such as foam tubing. Rigid bedrails can result in additional injury for active sleepers.</p> <p>For persons with HD who are likely to roll out of bed, the following suggestions may help to prevent injury from falling:</p> <ul style="list-style-type: none"> <li>• Place mats on the floor beside the bed.</li> <li>• Keep the bed in the lowest possible position.</li> <li>• Consider placing the mattress on the floor or, using a low bed frame.</li> <li>• Consider padding the edge of nearby furniture, such as dressers and headboards, with adhesive foam to prevent injury. This foam can be found in medical supply catalogs or at the local hardware store in the plumbing section.</li> </ul> <p>To address concerns about the person with HD getting out of bed alone when he/she is not safe to move without help, consider these options:</p> <ul style="list-style-type: none"> <li>• Bed or chair alarm systems are available for purchase or rental, such as through Patterson Medical (<a href="http://www.pattersonmedical.com/">http://www.pattersonmedical.com/</a>).</li> <li>• Use a remote or digital video "baby" monitoring system or a home security system.</li> <li>• Multiple applications, or "apps" now exist that allow tablets and iPads or smartphones to be turned into home monitoring systems.</li> <li>• In a facility, consider having the person with HD's room near the nurses' station.</li> </ul> |
| Late-Stage     | The Huntington's bed is a recently released product from Novacorr Healthcare: <a href="http://www.novacorr.com.au/product/huntingtons-bed/">http://www.novacorr.com.au/product/huntingtons-bed/</a> . This may be an excellent product for persons with HD who are very active but also unsafe to move without help.   |



## Social Participation/Leisure

|             |  |
|-------------|--|
| Stage of HD | <p><i>General Notes:</i> Socializing, participating in family or group activities, and being able to do leisure activities are meaningful for many people throughout the different stages of life, and these activities can be curbed by physical limitations, psychological concerns, and/or difficulty thinking. It is important to consider how to adjust social and leisure activities for greatest participation, as well as for enjoyment. With the progression of symptoms, the person with HD may find less enjoyment in activities that they previously liked and may find enjoyment in new or different activities. <b>Do the activities that can be enjoyed now, NOW! If you can enjoy travelling, go now! If you like to do family hiking trips and can enjoy doing that now, go now!</b> It is most important that the person with HD be able to feel that they are successful in these activities. Enjoyment can be lost when activities are too overwhelming; Occupational therapists call this idea finding the “just-right challenge.”</p> <p>Another resource for exploring “just-right” activities is by working with a recreational therapist, who is a professional that specializes in making “fun” accessible.</p>  |
| Early-Stage | <p>For someone who enjoys reading but is beginning to have difficulty with attention and memory, encourage them to <b>try reading short-stories, magazines, and newspapers</b>. The shorter length can reduce the burden of reading that often becomes frustrating.</p> <p>For someone who enjoys biking but is beginning to have difficulty with balance while riding, have them try using a bike with <b>wider tires and a step-through frame</b>. If balance is mildly decreased, it may be more helpful to consider a <b>recumbent bike</b>. For someone who has difficulty focusing and/or reacting quickly while biking, have them <b>ride in areas without vehicular traffic and to ride with another biker</b>.</p> <p>For someone who enjoys being an outdoorsman but is beginning to have some difficulty with focusing and remembering and/or balance, it is critically important that any <b>hunting activities be closely supervised and extreme caution</b> and attention given to gun safety. The use of tree stands is firmly discouraged. <b>Fishing presents less risk</b> in general and is an activity that usually can be more safely maintained. Supervision is recommended when fishing from a boat because of possible safety concerns.</p> <p>Some people with mild attention and memory issues find it more difficult to socialize on the phone, and they sometimes find it <b>easier to socialize through e-mail</b>. E-mail allows the person to read and re-read the communication at her own pace and when it is convenient.</p> <p>Some persons with HD have increased self-consciousness or anxiety in groups, which can make it harder to enjoy social activities because of concern about what other people think about their HD symptoms. It is important for family, friends, and caregivers to choose social activities or groups that are enjoyable by listening to any concerns and by recognizing anxious behavior. It can be helpful to socialize in smaller groups or socialize most with familiar people.</p> |
| Mid-Stage   | <p>Often with mid-stage HD symptoms, it becomes important to try similar but simplified versions of prior hobbies or to explore other hobbies. <b>It can be disappointing for people to try to do an activity that they were previously “good at”</b> and more enjoyable to try something new that they are currently “good at.” Some people may enjoy doing puzzles (depending on their level of chorea), playing cards (using a card holder), playing with pets, or tending to a container garden.</p> <p>Many persons with mid-stage HD find it more difficult to focus on an entire movie, unless it is a movie they already know, because it can be more difficult to remember the characters and the plot. It can be more enjoyable to watch sports (which can have new, interesting plays and a score board) or watch shorter TV shows.</p>   |
| Mid-Stage   | <p>In a facility, it is helpful to have as much <b>consistency</b> in staff and nearby residents and routine in order to reduce anxiety in this community-style residence. This can also help with supporting positive behavior and interactions.</p> <p>With possible increased anxiety of being around groups of persons during mid-stage HD, <b>social plans should be discussed and planned well in advance, as well as written on a visible calendar</b>.</p>   |
| Late-Stage  | <p>Socializing for persons with late-stage HD is often best enjoyed with <b>close friends or family members</b>. “Surprise visits” can be challenging and maybe less enjoyable. Ask friends and family to plan ahead and remind their loved one with HD to expect the visit.</p> <p>Leisure during late-stage HD may include spending time with close family or friends; looking out at a yard with birds, squirrels, etc.; looking at pictures; or looking at the TV (but not really paying attention to the content).</p>  |





A common component of most of the occupations discussed above is mobility, which includes walking or using assistive equipment to help with moving between places or changing positions. **Occupational therapists can perform assessments and provide recommendations to help persons with HD who have difficulty with mobility while performing occupations.** Recommendations for equipment can vary greatly depending on the progression of physical symptoms, such as chorea and decreased balance, and thinking abilities. Some persons with HD have a lot of chorea but their balance is only a little impaired. For others, balance may be horrible but their chorea is minimal. Chorea includes the spontaneous, unintentional movements that can be large movements, whereas balance involves the ability to avoid falling.

Many persons with HD have gradually worsening balance and chorea. The OT may recommend a **gait belt**, which is a wide belt that fits snugly around the person with HD's torso and a family member or caregiver holds it from behind (it should not be placed through belt loops). The gait belt is helpful for steadying a person with HD who is losing his balance in order to reduce the chance of an injury from falling. The gait belt is not intended to try to help reduce the chorea.

Another device that is commonly recommended for persons with mid-stage HD with impaired balance is a rolling walker with a seat, which is also known as a **rollator**. The rollator typically has four wheels, a seat, and hand brakes, and it is often more accommodating for chorea and balance problems. The rollator is also typically better for someone with thinking problems who would likely lift a **front-wheeled walker** and carry it, especially while turning. In general, front wheeled walkers are better for people who have very little (or no) chorea but still need a device to help with their balance. These assistive devices are tools for safely moving in order to perform activities of daily living. It is possible that devices may be needed for some activities but not for others. Sometimes, a person may need to use the rollator while working in the kitchen but may not need it to walk to the bathroom. *In essence, the person with HD may require different tools for different activities of living.* Please also see the chapter on physical therapy for further advice on ambulation and appropriate assistive devices.

Some persons with HD can become very unsteady as their symptoms progress which leads to a high risk of falling and injury. If this is a significant concern, the occupational therapist may recommend that the person with HD wear **hip bolsters (pads) and/or a helmet.**

When, or if, it becomes time for a person to be assessed for a wheelchair or specialty seating equipment, this assessment is often performed by an OT. A **Seating Evaluation or Wheelchair Evaluation** can be part of outpatient occupational therapy services, home health occupational therapy services, or take place in a Wheelchair and Seating Clinic. Wheelchair and Seating Clinics are often affiliated with inpatient rehabilitation hospitals and/or larger health systems. Therapists who work in Wheelchair and Seating Clinics are highly specialized in assessing a person's position, determining the most appropriate type of wheelchair or seating equipment, working with the durable medical equipment (DME) companies, and processing the paperwork needed by insurance.

A wheelchair fitting may include:

- A lower seat height (to make it easier to move the wheelchair by using their feet)
- Hill-holders (to keep the wheelchair from rolling backwards when the person sits down)
- Anti-tip bars (to keep the wheelchair from tipping backwards)
- A specialty seat cushion
- An adjusted seat angle
- A reclining back
- A lap tray/buddy
- A positioning belt
- A curved seat back with or without lateral (side) supports





For some persons with HD, a customized wheelchair does not provide enough support, and the therapist may recommend a different specialty chair. A **Broda® chair** is a commonly recommended specialty chair that provides a lot of support and padding for a person with HD. Some Broda® chairs fully recline.

Occupational therapists who specialize in helping persons with HD and their families and caregivers are a part of the healthcare team at HDSA Centers of Excellence. Occupational therapy is also available in local outpatient clinics, home health therapy agencies, rehabilitation centers and hospitals. If you need assistance finding help, contact HDSA by phone, email or use the locator button on the HDSA website.

## Resources

Huntington's Disease Society of America. (2015). Stages of HD. Retrieved from <http://hdsa.org/what-is-hd/#stages>

U.S. Bureau of Labor Statistics. (2014, May). Occupational Employment Statistics.

Retrieved from <http://www.bls.gov/oes/current/oes291122.htm>

Caregiver corner webinar

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/ptot-hd/>

HDSA Family Guide Series- Physical and Occupational Therapy-HD

<http://hdsa.org/shop/publications/#physical-occupational-therapy>

For more information on OT, please visit [www.aota.org](http://www.aota.org)





# Chapter 5

## Physical Therapy when **Caring** for a **Person** with Huntington's Disease



# Physical Therapy when Caregiving for a Person with HD

*Do you have questions or concerns on how to help your person with HD...*

*With walking, toileting or how to get in and out of a car?*

*Find the best walker and wheelchair for a person with HD?*

*Appropriate exercises for those with HD?*

*Please keep reading this chapter on how a Physical Therapist can assist your person with HD.*

## Impact of HD on a Person's Function

Movement problems lead to activity limitations and falls in persons with Huntington's disease (HD). Movement problems include:

- Bradykinesia (slowness of movement)
- Akinesia (delayed start of movement)
- Uncoordinated movement
- Motor impersistence (inability to sustain a movement like holding a cup in the hand without dropping it)
- Difficulty with rapidly alternating movements (turning the palm of the hand face up then face down rapidly)
- Difficulties performing sequences of movements (getting out of bed, standing up from a chair).

There are also involuntary movements such as:

- Chorea (brief, irregular movements): Chorea is typically seen first in the fingers, hands, and face muscles, and progresses to include all four limbs and the trunk.
- Dystonia (abnormal, sustained positioning of a part of the body): The most common types of dystonia are twisting of the arm with hand clenching, excessive bending of the knee, and twisting of the foot.

Younger age of onset is related to relatively more dystonia and less chorea, whereas an older age at onset is related to more chorea and less dystonia. Although chorea and dystonia are leading symptoms of HD, they do not predict the level of functional disability. Difficulty with daily activities and work functions is best predicted by movement problems such as bradykinesia and incoordination as well as cognitive problems.

Walking is affected early in HD and typically includes slower walking speed, shorter and more variable step lengths, legs spread wide apart during walking, and increased trunk movement compared to healthy adults. Balance problems usually occur in the early to middle stages of the disease. Persons with HD have trouble catching themselves when bumped or otherwise disturbed and have trouble walking or standing with their feet close together such as when they have to walk down a row of seats in a theater. They also may have trouble balancing when it is dark or they have their eyes closed.

As the disease progresses, falls occur frequently and often while the person is performing multiple tasks at the same time. They also fall when stepping around or over objects on the floor. Stairs are particularly dangerous and many falls occur while climbing stairs. Many factors cause falls including balance problems, walking problems, difficulty sensing where objects are in relation to one another, difficulties with eye movements, cognitive problems (decreased attention and ability to do two tasks simultaneously), and possibly psychological changes (impulsiveness, poor judgment leading to unsafe behaviors). The person with HD is likely to fall if he/she talks to family members while walking or if he/she carries something while walking or when climbing stairs. Impulsiveness leads to falls on stairs as persons with HD often approach stairs and begin climbing without stopping or slowing down. Family members will say that they appear to move rapidly onto the staircase as though propelling themselves over the edge of the

stairs. The person with HD is then unable to safely manage the stairs due to movement and visual problems. Teaching the person with HD to “stop, hold the rail, step slowly” can help reduce impulsive behavior and allow them time to visually scan and optimally use remaining movement skills.

Balance and walking problems affect the person’s ability to perform tasks such as reaching for objects, getting up and down from a bed, chair, or toilet, stepping in and out of a bathtub, or going up and down stairs. Hand coordination is often affected and makes writing, dressing, personal grooming, and cutting food and using utensils for eating difficult. In advanced stages, most persons will require help with all activities of daily living, relying fully on caregiver or nursing care.

## **Assist with Getting On and Off of Chairs, Beds, and Toilets and In and Out of Cars**

### **Person with Early and Middle Stage HD who is Still Walking**

Persons with HD may have difficulty doing transfers such as standing up and sitting down from chairs, beds, or toilets and getting in and out of cars. One way for a person with HD to learn how to transfer safely is through lots of practice. Caregivers should cue the person to do the transfer correctly and should not allow errors to occur. Through repetitive practice of a better transfer technique, the person with HD develops new, safe ways of doing the transfer. It may take 2-3 weeks of constant reminders and practice to develop a new way to transfer and turn it into a habit. If it is not learned after this time, it may not be possible to change the transfer.

### **Sit to Stand and Stand to Sit Transfer**

Chairs, couches, beds and toilets can be quite difficult to safely stand up from and to sit down on. It is easiest to sit down on and stand up from surfaces that are higher and firmer. For easier transfers it is best if a person with HD sits on surfaces that are the same height as their knees. If the surface is relatively firm it is also easier to go from sitting to standing.

#### **FAMILY NOTE**

*If you own a chair or couch that has become quite soft you can make the surface firmer by purchasing a piece of dense foam from a craft store and sticking it under the cushion of the chair/couch. The foam is not clearly visible and so the chair/couch still looks nice but is a safer seating surface for the person with HD.*

Toilets can also be quite challenging. If possible have a grab bar installed or purchase a bedside commode to place over the toilet. If you open the lid on the toilet and remove the basin from the bedside commode the person can still use the toilet while using the support of the bedside commode to assist in sitting down on and rising up from the toilet. Toilet transfers can also be made safer by using a raised or elevated toilet seat that can be easily attached to the toilet bowl without the use of tools.

People with HD often lean backwards and/or to the side while rising from chairs and tend to fall backwards into chairs.

A method that has been successful with many persons with HD is to teach them to transfer using their hands on their thighs. Instruct the person with HD to start rising from the chair with their hands on their knees. As they push through their hands for support they must lean forward and this puts their body over their feet. They then keep their hands on their legs sliding them up their thighs as they come to standing.

This provides for a smooth movement into standing up straight and steadies the body during the transfer. To return to sitting, a person places their hands on their thighs and slides their hands to their knees. This provides for a smooth and controlled movement to sitting and prevents them from falling backwards into the chair. By keeping their hands on their thighs, they are prevented from leaning backwards and thus do not tend to fall backwards into the chair. Caregivers may want to use cues such as “hands on knees, now up 1, 2, 3” using the count to cue a smooth



movement. If the person continues to fall backwards into chairs, a heavy chair with a wide base that does not tip back easily should be used.

Another common problem for a person with HD is that they start to sit down before they are close enough to the chair or bed and thus fall on the floor. Caregivers can help a person with HD to sit down safely by teaching them to always be sure that they are touching the chair with their hands or legs prior to sitting down. Caregivers may want to use cues such as “Touch-Turn-Sit” to help the person with HD to learn to safely sit down.

### Bed Transfers

Turning in bed can be difficult. It will be easier if the mattress is in good condition and relatively firm. It is also easier if bedding is kept simple and not tucked in tightly. Rolling side to side is easier if the legs are first crossed over in the direction of the roll and then the arms are used in a rocking motion to create momentum in the direction of the roll. For example, when rolling to the left side, the person with HD should first cross his right leg over his left leg. Next the person swings his arms side to side and finally to the left, creating momentum to help in rolling to the left. If chorea of the arms is a problem, turning in bed may be safer if persons with HD are instructed to grab their elbows and then swing their arms to the side to initiate turning. This prevents flailing movements that can lead to bumping the arms on the bed or hitting themselves. If chorea of the arms is impeding family helpers during a transfer have the person with HD “hug himself” to reduce arm chorea.

### Bed positioning and safety

Persons in middle to late stage HD may have problems with falling out of bed. This may be due to choreic movements that propel them out of bed, decreased spatial awareness that reduces their ability to sense the edge of the bed, and/or difficulties with force modulation causing them to “jump” out of bed when all they want to do is turn over or sit up. Possible solutions are to use padded bed rails to help keep them from accidentally propelling themselves out of the bed and to assist them in knowing where the edge of the bed is located. A lower bed height (3 to 5 inches from the floor) can improve safety and minimize injuries if the person with HD is still able to rise to standing from the bed. Another idea is to place a mattress on the floor beside the bed so if they fall out they won’t get hurt.

### Person in Late Stage HD who is no Longer Walking

When helping the person with late stage HD to transfer it is important for caregivers to use good body mechanics to prevent injury to themselves as well as the person with HD. Caregivers should place a gait belt around the waist of the person with HD (Fig. 1) and position themselves so that they are facing the person without any twisting or turning of their backs and with their feet spaced shoulder width apart. Caregivers may also place one foot in front of the other to make it easier to move forward and backward during transfers. Bending at the knees and grasping the gait belt, caregivers should ask the person with HD to help them and after a count of 3 to assist the person to stand up from the chair/bed/toilet or to sit down on the chair/bed/toilet. When a person with HD is no longer able to stand for transfers, a sliding board or transfer lift such as a Hoyer lift may be needed to safely transfer the person. Physical therapists can train caregivers on how to utilize these devices.



*Fig. 1: Standard gait belt that can be purchased for as little as \$8.*

### Bed Transfers

When helping a person with HD to get in and out of bed, let the person do as much as they can. To help them to scoot in the bed, have them place their feet on the mattress and lift their buttocks up and then move their hips up, down or to the side. Never move a person in bed by grabbing them under their arms and pulling. This can injure



their shoulders. It may be helpful to use a slide sheet to assist in moving a person with HD in the bed. To create a slide sheet take a twin-size sheet and fold it in half lengthwise and if needed also fold in half side to side. Place the folded sheet on the bed such that the person will be laying with this sheet under him from his lower shoulders to about mid-thigh level. Make sure the edges of the sheet extend away from his body to both sides. To move a person with HD up and down on the bed have a caregiver stand on each side of the bed and grasp the pull sheet close to the person's body. On a count of 3 lift the sheet and pull the person up, down or to one side of the bed.

### Bed positioning and safety

Persons with HD may continue to have problems with chorea leading to falls out of bed. If the use of side rails or other options are no longer working, a fully netted bed enclosure which fits over a standard hospital bed or twin bed may be helpful. Another option is a Craig bed® composed of a foam mattress and four padded walls that form a safe padded cubicle enclosure. While many persons may initially feel confined or isolated in these enclosures, they often appreciate the ability to change positions without falling out of bed and may sleep better. To reduce chorea and facilitate sleep it might be helpful to play soothing music that the person with HD enjoys or use weighted blankets that provide deep pressure to calm and relax the person.

### Getting into a car

When assisting a person with HD to get in and out of a car it may be helpful to place a piece of cloth on the car seat so that the person can slide more easily on the seat. A swivel cushion® may also be purchased (Fig. 2). If the person with HD is in a wheelchair, park the wheelchair as close as possible to the car. Open the car door as wide as it will go. Make sure the wheelchair brakes are on and the footplates have been removed or folded back. There should be enough room beside the car door so that the caregiver and person with HD can both move easily. Move the car seat back to give more room for the legs if needed. Ask the person with HD to slowly stand then step around so his back is towards the car. He should hold on to the car in the most secure but comfortable place. This may be the doorframe, the edge of the car seat or the edge of the dashboard. Support the person's body by using the gait belt as he starts to bend to sit down. To protect the person's head from bumping on the doorframe, place a hand gently over the person's head as he sits down. Ask the person to scoot back into the seat then help him to lift his legs over the doorsill. This method is much safer than if the person with HD tries to step into the car while balancing on one leg.



*Fig. 2 Swivel cushion and handy bar for car transfers.*

### Getting out of a car

Open the car door as wide as you can. Ask the person with HD to scoot towards the open door holding on to whatever is the most secure and comfortable surface. The person with HD should then swing both legs out of the car. Help to lift the person's legs out onto the ground if needed and have the person scoot forward in the car seat before standing. If the person with HD uses a wheelchair have the wheelchair ready to use with the brakes on. The person with HD can place his hands on the caregiver's upper arms or he may be able to place one hand on the door and the other on the caregiver's forearm for support if needed. The caregiver should not allow the person with HD to grab them around the shoulder and neck area as that may lead to the caregiver being pulled forward and off balance and could injure the caregiver's neck. Assist the person with HD to step away from the car or to step up to their walker. If the person with HD uses a wheelchair, have them step around towards the wheelchair so that their back is towards the wheelchair and sit down. Be sure that the wheelchair is placed close to the side of the car so that the caregiver can have the person with HD take 1-2 steps and then pivot and sit down in the chair.

## Helping to Walk

Person in early and middle stage HD who is still walking

Persons with HD should be encouraged to walk for as long as possible because it is important for their health and well being. Persons with HD who are inactive are at high risk for getting pneumonia and infections. Caregivers, in partnership with health care providers, should carefully weigh risk versus benefit in making recommendations to persons with HD regarding walking and when to transition to wheelchair use. Once someone begins to have falls during walking they are likely to continue to fall. Falls do not always lead to injury but it is important to realize that just because the first few falls did not result in an injury does not mean that the person will not be badly injured by falling at some time. If safety is an issue, consider the use of a walking device. The device that has been shown to be safest for use with persons with HD is the rollator or four-wheeled walker (Fig 3). Persons with HD may have difficulty using canes because they can't coordinate the use of the cane while trying to walk. Doing two things at once is often difficult for persons with HD and thus canes typically don't help and may make it harder for the person with HD to walk. Walkers with only two wheels in the front do not turn and can get stuck if the person with HD has chorea. Thus these walkers may cause a person with HD to lose their balance and make turning difficult and unsafe. Walking safety reminders include telling the person with HD to slow down when approaching obstacles, to focus their attention on walking, and to not walk and talk or carry objects like a gallon of milk at the same time. Try to make sure that walking paths in the home are clear of small objects and tripping hazards. When possible move furniture to make walking paths wider and more open. If the caregiver is giving physical support to the person with HD while he walks the use a gait belt is recommended for safety (Fig. 1 on page 60).



Fig. 3: Rollator Walker

The gait belt is placed on the person with HD at waist level and fastened snugly. This provides a safe and comfortable place for the caregiver to hold on. It is also easier to catch a person who is falling and possibly prevent a fall when using a gait belt. Avoid holding the person by the arm or under the arm as this can lead to shoulder injuries. Walking 3 times per day until mildly tired is an appropriate quantity to prevent further decline in health and function.

Footwear can lead to poor walking patterns and falls. The ideal footwear provides support to the foot and does not have a high heel. Shoes with a low heel can be worn as long as the heel is broad and provides a large area of support for the heel of the foot. Shoes should have contact all the way around the foot to ensure that the heel of the foot cannot slide off of the shoe. Mules, backless sandals and scuff slippers are not recommended.

John, a 55 year old male with a 12 year history of HD symptoms, is experiencing 3-4 falls every day, mostly when he is walking. He has had falls when turning while walking. He stated that when he holds on to his wife's arm or pushes a shopping cart he feels steadier. His physical therapist instructed him on the safe and proper use of a rollator walker. He walks more smoothly, faster and more safely with the walker. The walker was so beneficial that three months after using the rollator walker, John reported that he had no falls.

Person with Late Stage HD who is No Longer Walking Independently

If walking with the assistive device is no longer possible, use of a PVC Ambulator Walker or a Merry Walker can prevent inactivity and maintain leg strength and weight bearing for transfers (Fig 4 and 5). These types of walkers are large and require open spaces for use so they may not be feasible for use in private homes.



Fig. 4: PVC ambulatory walker



Fig. 5: Merry Walker®

## Walking on Stairs

Climbing stairs is a task that can be quite difficult for a person with HD. Stairs can be a safety hazard as a person with HD is more likely to have an injury if they fall on stairs. Ideally, use of stairs should be reduced or avoided. When stairs are a necessity, ensure that handrails are present. A person with HD may tend to move too fast and forget to hold the handrail. Caregivers can help by reminding them to:

1. “Stop, think about climbing the stairs”
2. “Grab the handrail” and then,
3. “Go slowly.”

Lisa, a person with HD, has had several falls when climbing stairs. She and her family are instructed that she is to stop before going up or down stairs and think about how she will climb the stairs. She is then to grab the hand rail and go up or down the stairs slowly. She is also told to be sure that she does not carry anything when climbing stairs and she is not to talk while on the stairs. Family is also instructed that they should not talk to her while she is on the stairs. They are also asked to cue her to “stop and think” before she goes up or down stairs.

## Protective Techniques (helmets, knee and elbow pads)

A person with HD at high fall risk, even when sitting or lying down, needs to wear protective gear to help reduce or prevent injuries. Soft helmets can protect the head from cuts and abrasions and provide some cushioning in the event of a fall. For a person who tends to land on their knees, pads can prevent abrasions and reduce bruising. Some persons bump their elbows on furniture due to their chorea and may benefit from elbow pads to protect them from injury. Some persons with HD tend to hit their heads when falling and are at lower risk for hip fracture than the elderly. If a person with HD is falling and lands on his/her buttocks, hip protector pads can greatly reduce the risk of fracture.

## Managing Cognitive and Behavioral Issues

Cognitive problems in persons with HD occur early in the disease and include difficulty paying attention and trouble thinking through the steps of an activity. A person with HD can also have difficulty thinking through complex problems and finding the best solutions to the problem. They may have trouble planning and putting in order the actions that they need to do to complete a task in the most appropriate and safe manner. A person with HD may also report difficulty with “multi-tasking.”

To improve learning, caregivers can:

- Encourage repetitive practice of tasks
- Allow ample time for the person with HD to process information
- Provide cueing such as reminding them of the next step or writing out a list of steps to be followed.
- Break down complex tasks into simpler tasks and have the person with HD attend to one task at a time.
- A quiet environment with few distractions can help the person with HD concentrate.

### Person with late stage HD

In later stages, the cognitive problems progress to dementia. These cognitive problems can interfere with the ability to learn new motor tasks. It is unlikely that a person in late stage HD can learn to do things in a new or different way. It will also be necessary to instruct them on each individual step of the task to be performed. Remember that a person with HD needs more time to process instructions, and due to bradykinesia, are slow to respond. When you give them an instruction allow extra time for them to respond.

## Breathing Exercises

Breathing function is affected in persons with HD typically in the later stages of the disease. These problems can be prevented or reduced by working on breathing exercises beginning early in the disease process. Breathing exercises such as pursed lip breathing (“blow out a candle”) and diaphragmatic breathing, as well as postural exercises are recommended at all stages of the disease to make breathing and coughing less effortful and to prevent lung infections.

- Pursed Lip Breathing is a way to forcefully blow air out that helps to improve strength of muscles used in breathing and keep the lungs clear.
  - o Tell the person with HD to relax his/her neck and shoulder muscles.
  - o Ask the person with HD to breathe in for two seconds, preferably through his nose, keeping the mouth closed. If the person cannot follow directions to breathe in through the nose it is okay if they breathe through the mouth.
  - o Then tell the person to blow out for four seconds through pursed lips. You can cue them by telling them to blow as if they are “blowing a candle out” or simply have the person breathe out twice as long as they breathes in.
- Diaphragmatic (Abdominal/Stomach) Breathing

The diaphragm is the main muscle of breathing. It normally does most of the work. This technique is best used when the person is feeling rested and relaxed, and while sitting back or lying down.

- o Tell the person with HD to relax the shoulders.
- o Place your hands on either side of his rib cage at the bottom of the rib cage.
- o Ask the person with HD to breathe in while focusing on expanding their ribs into your hands. As the person breathes in, the stomach should move outward. The person’s stomach should move more than the chest.



- o Then have the person blow out forcefully and slowly concentrating on bringing their stomach down and in while breathing out.

### Person with late stage HD who is no longer walking

At later stages, manually assisted coughing techniques (Fig. 6) or use of a mechanical Insufflator-Exsufflator machine may be helpful to clear lung secretions due to an ineffective ability to cough (Fig. 7). An assisted coughing technique is usually done while a person is sitting up but can also be done when lying. If the person is in a wheelchair, be sure to put the brakes on. The caregiver can stand behind the wheelchair and put their arms around the person with HD linking their hands together in front over the person's lower rib cage and upper abdomen (Fig. 6, left side). As the person with HD attempts to cough the caregiver pulls their hands up and inwards to assist the person. The caregiver can also assist coughing by positioning themselves in front of the person with HD and placing their hands over the person's lower ribs/upper abdomen (Fig. 6, right side). The caregiver keeps their elbows straight. As the person with HD attempts to cough, the caregiver pushes upwards and inwards. It may take practice to coordinate the cough with the motion.



Fig. 6: Assisted Coughing Techniques. From <http://www.spinalinjurycentre.org.uk/information/pdfs/019.pdf>.



Fig. 7: Cough Assist or Mechanical insufflator - exsufflator. From <http://www.spinalinjurycentre.org.uk/information/pdfs/019.pdf>.

## Wheelchairs and Seating Recommendations

### Person with Late Stage HD who is no longer walking

Seating can be a challenge for a person who has chorea and who adopts poor postures. A person with HD often likes to sit with their legs over armrests or leaning off to one side. While these postures can lead to contractures and other issues, it is difficult to address them therapeutically if the person is determined to sit that way and unable to understand or remember instructions to adopt a better posture. Placing a person with HD into a posture and restricting them from changing position can lead to agitation and combativeness.

When a person is no longer able to walk independently, they can often maintain their mobility by learning to self-propel using their legs while seated in a hemi-height or drop seat wheelchair (Fig 8). Standard



Fig 8 Standard height wheelchair and drop seat or hemi-height wheelchair from <http://www.rolandpriestley.com.au/SiteMedia/W3SVC257/Uploads/Images/Wheelchair%20Drop%20Seat%20Cushion.jpg>

wheelchairs (either self-propelling or transit) can be modified with a small wedge cushion or lumbar roll to promote better sitting posture, and padding for armrests and footrests to prevent bruising. Reclining wheelchairs provide the option to change positions from upright, but may encourage a person to slide down in the chair. Specialized seating including side supports and tilt-in-space seating may be necessary to improve upright posture of the trunk and prevent sliding out of the chair. For persons who do not tolerate this type of seating it may be necessary to add padding or use specialized chairs such as the Broda Chair® or the CareFoam™ chair (Figure 9). The CareFoam™ chair and others like it are particularly helpful with persons who tend to turn sideways in their chair and are best used in the later stages of the disease when self-propulsion is no longer possible.



Figure 9. Specialized chairs for HD

Broda® chair



CareFoam™ chair

## Staying Active

It can be difficult for a person with HD to stay active due to psychological symptoms of HD including depression, apathy (difficulty doing things even if they are interested in doing them), getting stuck on one thing or having difficulty shifting to new tasks, and impulsivity. Persons with HD who are depressed and/or apathetic may not be motivated to take part in or have difficulty starting an exercise program or physical activity. Many times apathetic persons with HD will take part in exercise if someone just helps them get started and works along with them to keep them interested. To help a person with HD become more active, try to find ways to be involved in activities that they have always enjoyed. It may be difficult for them to say what they would enjoy doing at the present time. What did they enjoy doing before they became symptomatic? Start here. If they liked dancing, then activities involving moving to music or dance would most likely be enjoyable to them. If they enjoyed reading, maybe they would like to listen to a book on tape while riding a stationary bike. There are many barriers that can make it difficult for a person with HD to remain active. Caregivers can ask themselves and the person with HD the following questions and then try to find solutions for any problems that they identify.

1. Do they need transportation to or from the activity?
2. Are they embarrassed to do the activity in front of other persons because of their HD symptoms?
3. Are they unable to physically do the activity? Could they do it in a new or modified way? For example, the typical way of knitting may be too difficult for some persons to perform but they may be able to do an easier type of knitting called arm knitting. Square dancing may be too difficult for a person to perform since it involves keeping up with the other dancers with fast movements but line dancing may allow a person with HD to modify the dance to their ability levels.

Setting a specific time during the day for a person with HD to exercise, and incorporating exercise into their daily routines, is a way to ensure they will do the activity/exercise. Encourage a person with HD to keep a log or diary of their daily activities or exercises so that they can see the progress they are making over time and reward them with



something they like for successfully completing their activity or exercise goals (e.g., being able to stand up from a chair without help or being able to stand on one leg to put shoes on) may help them to stay motivated.

For a person with perseverative problems (e.g. uncontrolled repetition of a behavior such as excessive hand washing), try to redirect their attention to other tasks. It is always best to give a person with HD specific time limits for exercise programs to avoid having them over exercise due to perseveration. Impulsivity and lack of awareness of deficits in persons with HD can result in unsafe behaviors. Talking with a person with HD about the “pros” and “cons” of their behaviors, and providing constructive ideas of other ways to behave may be helpful.

### What type of Exercise/Activity is best for persons with HD?

Research studies have suggested that exercise is beneficial for reducing symptoms and maximizing function in persons with HD. Maintaining a healthy heart is important to the health of all persons, no matter their disability. Persons with HD should engage in aerobic activities ideally for at least 150 minutes a week. Walking is a good aerobic exercise and still possible for the person in the middle stage of HD to perform. If available, stationary bikes are an excellent means of providing aerobic activity. Care should be taken with bikes that require the rider to step over a bar and lift themselves up to the seat. A person with HD is often unable to balance on one leg to step over the bar and so is likely to fall if asked to mount this type of bike. It is easier to climb on and off a bike with a very low center bar and a seat at thigh height or a reclining bike with a standard chair seat that swivels.

A person with HD may develop muscle and joint contractures (abnormal shortening of muscles that restrict joint movement) despite their choreic movements. To prevent contractures, active range of motion (AROM) exercises in which the person with HD moves arm and leg joints through their full range of motion should be performed daily and should focus on straightening the joints. AROM exercises should be done while the person with HD is lying down or while seated in a supportive chair so that the person can maintain their balance with minimal effort while attempting to move their arms and legs to the ends of movement at joints. By the middle and late stages, active movements are often limited to very small ranges of motion; thus caregivers may need to manually help the person move through the full joint range of motion. Caregivers should not force movement of a joint; instead they should gently and slowly move the joint through its full range. If the person with HD says a movement hurts then stop the movement before causing pain.

A person with HD often becomes weaker in the middle stage of the disease. Activities or exercises to maintain strength should be provided. The use of weights is typically not feasible given other problems such as chorea and difficulty holding a muscle contraction. Chorea can cause a person with HD to strike themselves with weights or with weighted arms or legs. At this stage, a functional strength-training program is most appropriate. For leg strengthening, have the person with HD stand up and sit down from a firm, stable chair 10 times (or as many times as they can if the person cannot do 10 times). Repetitive sit to stand transfers can be quite beneficial both for increasing leg strength and improving function. Persons with HD have difficulty maintaining their weight and exercise programs can help to improve appetite but should be kept at a level that won't lead to even more weight loss. Functional activities such as lifting objects onto shelves help maintain arm strength. Having the person with HD push and pull a wheelchair or other wheeled object can help strengthen back and chest muscles. Adding weight to the wheelchair or wheeled object is a way to ensure that this activity provides enough resistance to be beneficial.

Balance problems that start early in the disease can make it difficult for a person with HD to stand with feet close together or balance on one foot and increases their fall risk. By the middle stage, excessive trunk movements can also put a person with HD off balance. Additionally persons with HD lack insight and therefore do not understand that they are at high fall risk and don't modify their movements to ensure their safety. To improve balance, have the person with HD stand in front of a counter with their feet together and try to balance without holding on to the counter. If the



person is able to do this then have them add head turns, arm movements up and down, or closing the eyes while holding the same position. These exercises can be further progressed by repeating the same exercises while the person stands on a soft surface such as foam or the person stands on one leg. The person can also practice walking forward and backward beside a counter placing one foot in front of or behind the other to challenge their balance.

#### EXERCISE AND FITNESS - MIDDLE STAGE HD

Rob lives at home with his wife with additional caregiver assistance during the days when his wife is at work. Rob is 40 years old and was diagnosed with Huntington's disease 15 years ago. He can ambulate but has fallen repeatedly in the last year. He walks with his legs spread in a wide base of support and tends to clasp his hands behind him. When getting up from a chair he requires the use of his hands and has to rock back and forth several times. He tends to lean backwards during the transfer and has fallen backwards sometimes landing on the chair and sometimes missing the chair and falling to the floor. When sitting down in chairs Rob stops several feet away from the chair then falls backwards into the chair with a stiff trunk. He does not bend at the hips and chairs tend to slide backwards when he lands in them. He has broken two chairs when sitting down in them, both times the chair tipped over backwards.

Training Rob to transfer by first feeling the chair on the back of his leg and then by placing his hands on his thighs to encourage him to bend forward and provide support throughout the transfer will improve safety during stand to sit transfers. Due to cognitive changes in the middle stage of HD, Rob will need to practice this new method of transferring many times before he will remember to do it. Visual and verbal cues are also helpful in reminding him to do the transfer in a safe manner. To further improve safety during transfers, chairs in Rob's home should be heavy so that they don't tip over when he falls into them. There should be grab bars in the bathroom so that he has a safe, firm handhold for toilet transfers and when bathing.

He may benefit from walking with a rollator walker. These walkers encourage users to keep their arms in front of their bodies and provide constant support. The swivel wheels will allow Rob to walk and turn smoothly without having to lift or maneuver the walker. Most persons with HD are able to learn to safely use the rollator with little to no instruction. The brakes are not necessary for safety but family and caregivers, as well as Rob, should be trained to lock them before having Rob sit on the seat of the walker.

If Rob has been sedentary he would benefit from a program to improve his activity level while increasing strength and improving balance. Walking programs are highly recommended. Due to apathy related to HD, short daily walks are usually better tolerated. Encourage Rob to take on some chores/household activities that will require him to get up and walk multiple times a day. Some suggestions are that he do things like getting the mail, check that the porch light is turned on, carry his dishes and trash to the kitchen using the basket on the rollator walker. The chores and activities should be individualized based on Rob's living environment and his interests. Since many persons with HD spend a large amount of time watching television it is recommended that persons with HD stand up and walk each time there is a break in the show for commercials. Rob could walk to the television, touch it and return to his chair or walk to the bathroom or any other appropriate target. Build in rewards that are individualized to help overcome the apathy that is present in HD.

#### Person with late stage HD who is no longer walking

It is beneficial for all persons with HD to remain active. In late stage HD this can be quite challenging due to difficulty moving, apathy and cognitive problems. Remember that apathy is a difficult thing for the person with HD to overcome and no amount of arguing will make them less apathetic. Find things for them to do that they are likely to enjoy and that are simple and easy to accomplish. It may be easiest to get them to engage in activity if it is a means to getting something they want. For example caregivers might have them self-propel their wheelchair with their feet to get to the kitchen to eat their dinner. If walking is no longer feasible then self-propulsion of the wheelchair should be encouraged. It is difficult for a person with HD to self-propel the wheelchair with their arms. It may be easiest for them to propel the chair by pushing it with their feet. Sometimes pushing it backwards is the easiest method. If this is feasible it is a way to allow them to remain active and experience some independence. Having a person with

HD reach for objects like a cup or the remote control if they are able, rather than just handing it to them, will help to keep their arm muscles strong.

#### LATE STAGE HD

Sarah is a 54 year old woman diagnosed with Huntington's disease 20 years ago who lives in a nursing home. She is dependent in all activities of daily living and is able to follow simple commands. Sarah used a rollator walker in the past for ambulation but is no longer able to safely use one due to her motor planning and balance deficits. She is able to rise to standing and can walk short distances (e.g. from her bed to bathroom) with minimal to moderate assistance from one person before fatiguing. The staff at the nursing home are concerned because she is falling out of bed and geri-chairs due to her severe chorea and frequently falls in her room when attempting to walk unassisted to the bathroom. Sarah is a good candidate for the use of the Broda® or Carefoam™ chair to prevent falls. She might also be able to take a short walk once or twice a day using a Merry Walker.

There are many things that can be accomplished to help a person with HD maneuver through their world. A Physical Therapist is an important person in the team of professionals to help keep your person with HD active, comfortable and safe.

## Resources

Caregiver Corner webinars

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/ptot-hd/>

Caregiver Corner — Gait and Balance

<https://www.youtube.com/watch?v=VJHc9nXQxCs>

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/gait-balance/>

Physical Therapy Guide

<http://hdsa.org/shop/publications/#physical-occupational-therapy>

Safety in the home

<https://www.youtube.com/watch?v=MwlJLvmusRc>



# Chapter 6

## The Role of Speech-Language Pathologists in Huntington's Disease



# The Role of Speech-Language Pathologists in Huntington's Disease

*Do you have concerns about your person with HD's swallowing?*

*Are you concerned about the difficulty the person with HD has following directions about eating?*

*Please keep reading about how a Speech-Language Pathologist can provide some important information on swallowing.*

## Introduction

The goal of this chapter is to provide caregivers with a background of knowledge relating to the challenges of speech/communication, swallowing and cognition to better understand and care for their loved one with HD. It is important for family members to share information across the different caregivers that a person with HD will encounter.

Speech-Language Pathologists are professionals who are trained to evaluate swallowing and determine the most appropriate diet for the person with HD with the goal to swallow safely, maintain nutrition and avoid aspiration. This evaluation, early in the disease, in a clinic setting typically begins with a swallow exam that includes evaluating how a person swallows, a motor speech examination and the swallowing assessment. This evaluation gives the team baseline information on how well the person with HD is currently functioning. Further evaluation can include a Modified Barium Swallow Study (MBSS), a test that takes place in the Radiology department. During this test, the person is seated in a chair and various textures of barium are swallowed to represent different food textures and liquid consistencies. This allows the Speech-Language Pathologist and the Radiologist to accurately evaluate the type and severity of swallowing difficulty (dysphagia,) and observe if there is any sign of aspiration. See the chapter on social services that discusses ways to pay for services.

## Communication Impairments and Huntington's Disease

At some point during the progression of the disease, persons with Huntington's disease will develop communication and cognitive impairments. Most commonly observed may be motor speech impairments. A motor speech impairment refers to speech disorders resulting from neurologic damage. Common *motor speech impairments* are:

Dysarthria: a motor speech disorder in which the muscles of the mouth, face and respiratory system become weak, move more slowly or not at all. Symptoms of dysarthria include: "slurred" speech; speaking softly or whispering; slowed rate of speech; rapid rate of speech or mumbling; reduced tongue, lip and jaw movement; abnormal intonation and rhythm; changes in nasality when speaking; hoarseness/breathiness; and drooling or poor saliva control.

Apraxia of Speech: Also commonly known as verbal apraxia, this motor speech disorder involves difficulty with sequencing sounds in syllables and words. Apraxia can be seen in conjunction with dysarthria. Signs/symptoms include approximations of words (knowing what they want to say but the word comes out wrong). For example, a person may want to say "table" but may instead say "biple." The brain has difficulty coordinating the muscle movements necessary to complete the word and therefore the person may try again, only to have a completely different word come out, such as "terple." Due to the variety of errors presented, it can be challenging to understand a person with apraxia of speech and very frustrating for the person with HD who is attempting to communicate and producing multiple errors.

Other communication challenges that persons with Huntington's disease face include difficulty with word finding, stuttering, reduced voice quality (hoarse, breathy or harsh) and poor coordination of breathing and speech which impacts voice quality and volume.





# Cognitive Changes in Persons with Huntington’s Disease:

Cognitive changes may make it difficult for persons with HD to live independently and care for themselves. This may present in the following ways:

- Memory impairment (immediate and short-term)
- Poor reasoning/judgment – lack of awareness of deficits, reduced safety awareness
- Reduced problem-solving ability
- Difficulty sequencing/organizing thoughts or ideas
- Easily distractible/short attention span
- Poor concentration
- Reduced ability to learn new things
- Problems with numbers/mathematic computation

These kinds of cognitive changes make it difficult for a person with HD to complete simple tasks throughout the day, and therefore present challenges related to their care. Learning to develop clear, appropriate communication strategies to assist the person through activities is imperative in allowing them to maintain independence for as long as possible.

Cognitive impairments can cause communication breakdown in a variety of ways. A person with HD may get “stuck” on certain words, have trouble taking a turn within a conversation and therefore may be seen as rude when they interrupt the conversation or change topics before allowing the caregiver to complete a statement or thought. It is important for others to recognize that the pattern of communication is not indicative of personality, but rather impairment, and take extra care to be patient and understanding.

**FAMILY NOTE**  
*Do not react negatively to what is being said. I’ve learned to say less and listen more. Many times my loved one is expressing a frustration that passes quickly. If you react, the frustration escalates to anger.*

### Cognitive Strategies:

Persons with HD may have difficulty learning new information. Some common strategies for cognition include:

- Problem Solving – Caregivers should speak with their loved one about how they might solve a simple problem.
- Orientation – Use a calendar (preferably large, visible) to help identify appointments and activities.
- Memory – It is important to establish a routine specific to the person.

**FAMILY NOTE**  
*A dry erase board was helpful to write dates, meals and appointments. When we are unable to understand her, we ask her to repeat her statement and repeat the statement back to her asking if that’s what she meant.*



## Tips to Improve Communication

Communication is how all human beings relay their thoughts, feelings, needs and wants to those around them. People with communication challenges are quite often left without a proper means to relay information. Here are some tools to try to optimize communication for all involved.

### Tips for the Listener:

- Reduce environmental distractions (TV, radio, cell phones, computers, close door).
- Ask Yes/No Questions – Do not use open-ended questions.
- Ask questions with only two or three options (e.g. “Do you want water or coffee?”)
- Allow adequate response time – Do NOT rush the speaker.
- Keep it simple! - Do not make communication too complex. Remember that cognitive impairments/memory may play a role.
- Have a schedule – Let the person know what’s expected. If memory is a problem, recognize that they may forget to do a task if it’s not on the schedule.
- Only ask ONE question at a time – Wait for the answer, THEN move to the next question.
- Do NOT pretend you know what was said! Ask clarification questions, rephrase, but do NOT give up. What they say is important and needs to be understood!

### Tips to give the Speaker/Person with HD:

- Encourage the person to speak clearly/exaggerate the sounds.
- Encourage the person to speak slightly louder (especially if volume is reduced).
- Encourage the person to take a deep breath before speaking.
- Ask them to speak one word at a time.
- Encourage them to use gestures to assist with communicating.
- If he/she can’t think of the word, ask to “describe it.”

#### *FAMILY NOTE*

*I always talk to him about how things are going, even if he can’t respond.*

## Augmentative and Alternative Communication

When persons with HD have significant verbal communication impairments an SLP can look into the use of another system of communication called Augmentative and Alternative Communication, or AAC. There are many types of AAC systems, from pointing and gesturing, to blinking and high-tech computers. Two basic types of aided communication systems can be considered for those with HD; a static communication board and a dynamic display/speech generating device.

Static Display – Static refers to items that are fixed to a particular location and do not change. Frequently, static communication boards will involve photos of commonly used objects or needs related to daily care. The person with HD can either point to the photo or with some low-tech voice output devices a caregiver or the person can record a message for each button.

Dynamic Display Communication Devices - Dynamic display communication devices refer to devices in which the display changes to show other pages that may be linked by topic or other categorization techniques. Commonly,



dynamic display communication devices are computer based and have synthesized speech for verbal output. Dynamic display communication devices can allow the person to customize buttons or pages specific to their needs and can change easily depending on the need or environment.

It is important to recognize that not all persons with HD can successfully use an AAC system. If there is decline in function or inability to learn new information due to cognitive functioning, AAC may not be appropriate. If a person receives a high-tech computerized speech device, many people involved in the care of the person may need to be trained on that specific system. A low-tech option like a static communication board, including an alphabet board for spelling and picture/symbol based options representing common items associated with activities of daily living, may be most appropriate and easily used by all involved in the person's care.

## Dysphagia

The term dysphagia refers to difficulty swallowing. Dysphagia is very common in persons with HD, and is a major contributor to aspiration pneumonias and weight loss in persons in later stage HD. It is vital that all involved in the persons' care are aware of signs and symptoms of dysphagia, as well as ways to reduce the risk of aspiration. Laryngeal aspiration — material entering the airway and passing to the lungs — can lead to pulmonary infection or pneumonia.

There are multiple phases to the swallow mechanism, and persons with HD can have dysphagia in any or all of the phases. Below are descriptions of the phases related to swallow and symptoms of dysphagia associated with these phases.

Oral Phase – Involves moving food and liquid through the mouth and preparing the mass of food or liquid (bolus) to be swallowed. Persons with HD may notice difficulty chewing or moving the food or liquid within the mouth. The tongue, lips and jaw may also be uncoordinated (part of the movement disorder symptoms) and therefore result in food being held in the mouth. Persons with HD may also take too large a bite. This is worsened with impulsivity. With oral dysphagia, you may also see food or liquid falling out of the mouth.

Pharyngeal Phase – food or liquid passes through the throat (pharynx). The actual swallow may be delayed resulting in food or liquid spilling into the throat before the person is ready to swallow. There may be weakness of the muscles of the throat resulting in food or liquid sticking in the throat or spilling into the trachea or wind pipe. The action of food or liquid entering the trachea is called aspiration. A possible indicator of food or liquid reaching the wind pipe is a wet voice or coughing before, during, or after the swallow.



The severity of dysphagia increases as HD progresses. Symptoms will progressively worsen and a persons' choreic movements may make it difficult for them to feed themselves or to be fed by others.

## Signs/Symptoms of Aspiration

As mentioned, an early sign of difficulty swallowing will be a cough or throat clearing. It is important to give detailed information to the physician or the Speech-Language Pathologist so they can develop an appropriate treatment plan. Does coughing or choking occur once or more per meal? Once per day? Only when in a reclined position? Only with liquids? Only with certain foods? The cough is the body's way of removing any material from the airway. Warning signs of dysphagia and/or aspiration include:

- Increased coughing episodes, especially immediately after swallowing.
- Wet sounding voice.
- Slower rate of eating or drinking.

## When to Consult with a Healthcare Provider:

- Wet sounding breathing.
- Frequent congestion.
- Fever, clear evidence of an upper respiratory infection.
- Continued weight loss.
- Choking episodes.

As symptoms of HD progress, a person's hand to mouth coordination may deteriorate and make it difficult for them to feed themselves. Modifications can be made to their food, such as offering more finger foods or preparing foods that are softer and easier to pick up with a spoon or fork. An Occupational Therapist can recommend adaptive or assistive equipment such as a built up handle for utensils or a mat that stabilizes a plate or bowl. A sippy cup with a straw may help to prevent spillage when drinking liquids. The use of a straw can also limit the size of the sip, thus making swallowing safer. It's important to encourage the person with HD to maintain a neutral head position when drinking liquids rather than throwing their head back to drain the liquid from the can, bottle or cup.

### FAMILY NOTE

*Do your best to try to make your person with HD feel that they are still helping in their own care.*

The person with HD who has dysphagia is also in danger of not getting enough calories during a single meal or across the day, if they struggle with spilling food either from the utensil or from their mouth. Eating more, smaller meals during the day can help add calories. Caregivers may be reluctant to assist the person with HD and most persons with HD want to maintain their independence and may not willingly accept help. Reminders about proper positioning while eating and about taking small bites and sips may need to be regularly repeated.

### Behaviors:

**Impulsivity** – Some persons with HD stuff food in their mouth and continue to do so without obvious or visible swallowing. This could lead to a serious episode of choking. The sequence of bite-chew-swallow before the next bite may be difficult to get across as cognitive decline continues. Verbal reminders or a light touch on the person's arm may be good tactics to prevent impulsivity. Caution with liquids is also very important. Persons with HD may have a tendency to guzzle or gulp liquids without stopping for a breath. One sip at a time may be difficult to "enforce" but is a strategy that could work, especially if the person is experiencing coughing/choking during or after the swallow, or if they demonstrate a wet, gurgly voice quality when they're done with their beverage. Adult "Sippy" cups or a straw may be an option.

**Excessive sleeping** may impact the ability to meet caloric needs as meals are missed. It's important to encourage a regular schedule of meals and snacks.

### FAMILY NOTE

*Let her develop her own sleep pattern, even if it is out of sync with the rest of the world.*



Write out a schedule on sticky notes, or on the calendar in the kitchen. For example:

- **Lunch at Noon – sandwich/pudding/milk are in the fridge**
- **Snack at 2pm – ice cream bar in the freezer or Ensure® in the fridge**

Consult the person's medical provider for possible sleep aids to help regulate sleeping patterns to allow for more daytime wakefulness.

**Food refusal** is a behavior that may be related to the person's realization that the texture may be difficult to chew or swallow. Perhaps they had a previous choking episode related to that type of food. Food likes and dislikes may vary as the disease progresses or may vary from one day to the next for no discernable reason. Medication side effects may also affect how food tastes.

**Excessive consumption** of anything can be problematic (caffeine, smoking, alcohol). For example: During a recent clinic visit, a person reported that she typically drinks 2-3 diet sodas per day while her sister held up 6 fingers, indicating the actual number. Discussions around excessive consumption can become problematic when a person shows poor judgment or is unaware of their excesses. Some families have coped with this behavior in a variety of ways, including a lock on the refrigerator or freezer, setting up a rigid schedule for the next caffeinated beverage throughout the day, or creating a reward system for proper eating.

#### *FAMILY NOTE*

*One survey respondent said "no caffeine after noon or try to eliminate it altogether."*

**Dangerous food texture choices** – While the person's chewing/swallowing coordination deteriorates even further, their judgment is also declining. Even though caregivers may carefully prepare balanced meals with texture modifications, the person's snack choices may not be aligned. Here are examples of food textures that create "red flags."

**Particulate food:** Nuts, popcorn, trail mix, dry rice, peas, corn. Placing a few nuts in the mouth at one time and chewing immediately breaks them into small particles – similar to placing a spoonful of rice, peas or corn in the mouth. If there is any tongue or jaw weakness, this texture will be very challenging. While the person is chewing and coordinating the small particles in the mouth, a small piece could fall into the airway causing a choking episode while particles are still in the mouth.

**Dense, chewy food:** Meats like pork chops and steak, dense breads or bagels. Without intact chewing and coordination, this texture is especially challenging and often the first texture to require modification (i.e., chopping or grinding) or to be eliminated from the diet altogether.

**Dry, hard, crunchy food:** Crackers, chips, raw carrots, apples. These types of foods like the particulates can also break into little pieces making coordination in the mouth difficult. The dry texture can make it difficult to move the bolus from the front of the mouth to the back where it can be swallowed. Soft, canned fruit and cooked vegetables are ways to provide important nutrients in a more manageable way.


**Stringy food:** Lettuce, celery, pea pods. With these textures there's a sensation that it sticks in the throat making it difficult to swallow. It's easiest to eliminate these textures from the diet and substitute cooked vegetables as mentioned above.

**Some candy:** Candy can be difficult to chew and/or swallow. Textures such as nuts and caramel can combine to create a challenging combination to chew. Choose softer, easy to chew candy bars or pieces of candy. Avoid types of candy that are sticky, crunchy, hard or chewy.

#### *FAMILY NOTE*

*I always tried to let my husband lead as normal a life as possible. This perhaps put him at risk of choking or falling a little more but I thought the quality of his life was more important than the quantity.*





**Holding food in the mouth** – This can be problematic not only for the person with HD, but also for the caregiver, especially if they're involved in feeding the person. A gentle reminder to swallow, and careful observation of the person's larynx (Adam's apple) to see the rise and fall that indicates the person has swallowed, will help to determine when it's safe to present the next bite or sip.

Developing strategies for modifying the texture of solid foods, and considering the possibility that liquids may be safer if thickened, will be vitally important as HD progresses. The change in swallow function happens gradually over time and family members may have witnessed a similar decline in other family members.

A common scenario is that although the care provider is aware of changes that need to be made to food texture, the person adamantly refuses the changes – unaware of any difficulty or simply not wanting to be singled out as different.

Some strategies for the person with HD at meal times include:

- Sit upright at the table for all oral intake including medications. Avoid sitting in an easy chair in front of the TV.
- Avoid distractions when food or liquid is in the mouth. Caregivers should wait to ask a question until the person has swallowed.
- Encourage small bites and single sips. Remind the person to maintain a neutral head position instead of throwing the head back to drain the liquid from the cup. Introduce the use of a straw if the person seems to be coughing when drinking from a cup.
- Encourage a second swallow to clear food from the mouth.
- Have the person with HD take a sip of liquid frequently throughout the meal to clear any residue in the mouth.
- For medications: Advise the person with HD to take one pill at a time with a sip of liquid. If coughing occurs, try placing the pill in yogurt, applesauce or pudding. Crushing the pill and placing the powder in applesauce may also be a safe option.

#### **FAMILY NOTE**

*I was worried that my daughter was not getting her medications because she was having trouble swallowing liquids. The SLP helped us by recommending different cups that made it easier for her to drink liquids. The SLP also talked about food textures and ways for my daughter to be safe while swallowing.*

It is important to supplement what is eaten during meal times with snacks. Keep healthy snack options easily accessible such as single serving cups of diced peaches, pears, yogurt or applesauce. Other options such as sport shakes, individual cans of tomato/vegetable juice, smoothies, and string cheese can be stocked in the fridge for easy access. If weight loss is a concern, consider snacks higher in calories such as ice cream, milkshakes, pudding or full fat yogurt. Drinking water throughout the day is important to maintain hydration but it may be appropriate to substitute beverages with calories like fruit juice or milk (at least 2% or whole). Supplements, such as Ensure® or Boost®, may be recommended by the physician or dietitian.

When is it appropriate to thicken liquids? What are the implications? If the person is coughing with regular thin liquids, first try introducing a smaller sip or a single sip at a time or try a straw. Notice if there's a pattern that results in coughing like talking with liquid in the mouth, distracted drinking, too large of a swallow or reclined in a chair in the living room. Next try naturally thicker liquids such as tomato juice or a smoothie. Most physicians and speech-language pathologists agree that a formal swallowing study is indicated prior to thickening liquids on a "permanent" basis.

Aspiration, as mentioned earlier, is a real risk for persons with Huntington's disease. Coughing when eating or drinking may be a sign that the person is at risk for aspiration or that they are already aspirating. Sometimes coughing may





lead to the more serious condition – choking. We’ve all choked at some point. Have you ever experienced a sip of liquid or a crumb of food “going down the wrong pipe”? That’s a choking episode – you cough hard and a lot, you can’t breathe right for a few seconds, your eyes water, you might not be able to speak. As described above, the cough (or choking) is our body’s reaction to ridding the wind pipe of the offending liquid, food or foreign object. Thankfully, most of the times, people remain conscious, continue to cough and eventually recover from the choking episode.

If a piece of food lodges in the windpipe and coughing does not clear it out, this is much more serious. A choking person’s airway may be blocked so that not enough oxygen reaches the lungs. The person may become unconscious if the airway is blocked. Either conscious or unconscious, the person needs help quickly! It’s extremely important that caregivers living or working with persons with HD learn the Heimlich maneuver; an emergency technique to help clear someone’s airway. There are many how-to and YouTube videos available on the internet to help you learn.

Indications that may lead to an alternative feeding method such as a feeding tube occur when the person with HD continues to lose weight, is fearful of eating or drinking, or is malnourished or recently hospitalized for pneumonia. This is a discussion that the person with HD, along with their family members/caregivers, should ideally engage in early in the course of the disease. The physician, speech-language pathologist and/or dietitian are all professionals that can ask the questions and engage in discussion with the patient/family.

Getting assistance from a speech-language pathologist can help you, the caregiver, feel more comfortable keeping your person with HD safe while also considering quality of life issues related to speech and eating.

## **References**

HDSA’s Speech-Language and Swallowing Difficulties Guide is available for download  
<http://hdsa.org/shop/publications/#nutrition-swallowing>



# Chapter 7

## Nutrition

and

## Huntington's Disease



# Nutrition and Huntington's Disease

*Do you have questions about the kind of diet that might work best for your person with HD?*

*Do you have questions about thickening liquids and pureeing hard foods?*

*Have you talked with your person with HD about a feeding tube?*

*Please read this nutrition chapter to learn about these issues and others to consider.*

## Introduction

More people are coming to realize the importance of good nutrition in maintaining health and preventing disease. Advice has been issued by many organizations about diet and prevention of cardiovascular disease, cancer and other disorders. This advice focuses on reduction of calories, avoidance of *trans* fats, and increased consumption of fruits, vegetables, and whole grains. While these guidelines are appropriate for the general public, nutritional issues and eating with HD introduce additional considerations. Diet can not prevent or cure HD, but it can contribute to maintaining maximum functional ability and quality of life.

## At-risk and Early HD – a Brain Healthy Diet

### Nutritional Needs in Early Stage HD

Depression resulting from the diagnosis can cause decreased appetite and/or changes in usual eating habits. Some persons with HD have reported food cravings, especially for high carbohydrate foods. There is usually no harm in indulging these cravings, as long as other foods are eaten to maintain a balanced diet. However, any significant weight changes, whether an increase or decrease, should be reported to your health care provider.

### FAQs:

#### What kind of a diet should I follow?

Having a diet that is adequate in calories to maintain a healthy weight is the most important dietary factor with HD. Individualized diet plans are the best for the ever-changing needs of a person with HD.

#### Is there something I can eat or take to prevent or limit HD (i.e. a 'magic' food or nutrient)?

Research on nutrition specific to HD is very limited. Most recommendations are extrapolated from studies of other disorders, as well as those for general health. To date, no "magic" foods or nutrients have been discovered.

#### If I eat more of any specific food will that help my HD?

Based on research literature for the general population, and some more common neurological conditions, such as dementia, there are some dietary factors considered to be "brain healthy." Two of the most important are *anti-oxidants* and *anti-inflammatory agents*. There are many foods that provide these substances, so you can take your pick of these foods to incorporate into your diet. Try to include at least one at every meal.

#### - Healthy Fats

Some mono and poly-unsaturated fats have been found to have anti-inflammatory properties, and also been shown to be of benefit in a number of chronic degenerative diseases. Sources include: fatty fishes (such as salmon, mackerel and sardines), olives and olive oil, avocados, walnuts, and flax seed.



## - Antioxidants

A class of substances found in fruits and vegetables are called “phyto-nutrients.” Many of these are known to function as antioxidants. While no one specific antioxidant is recommended, a diet rich in a variety of fruits and vegetables is appropriate for anyone. Color is important when choosing foods with antioxidant properties. Foods with deep, rich color tend to have the highest amounts. The table below lists some examples of food sources of various phyto-nutrients.

### Color Code Sources of Phyto-nutrients in Vegetables and Fruits

| Color         | Phyto-nutrients                    | Sources   |
|---------------|------------------------------------|---|
| Red           | Lycopene                           | Tomatoes, pink grapefruit, watermelon, guava          |
| Purple/Red    | Anthocyanins, polyphenols          | Berries, grapes, red wine, prunes                     |
| Orange        | $\alpha$ -, $\beta$ -carotene      | Carrots, mangoes, pumpkin                             |
| Yellow/Orange | $\beta$ -cryptoxanthin, flavonoids | Cantaloupe, peaches, oranges, papaya, nectarines      |
| Green/Yellow  | Lutein, zeaxanthin                 | Spinach, avocado, honeydew, collard and turnip greens |
| Green         | Sulforaphanes, indoles             | Cabbage, broccoli, Brussels sprouts, cauliflower      |
| White/Green   | Allyl sulphides                    | Leeks, onion, garlic, chives                          |

Data from: Heber D. Vegetables, fruits and phytoestrogens in the prevention of diseases. *Prostgrad Med* 50:145, 2004.

### Should I continue to follow a low fat diet for my cholesterol (or other diet)?

If you have been advised to follow any therapeutic diet, you should continue it as instructed. At your next follow-up appointment, ask about the best way for you to incorporate more “brain healthy” foods into your diet plan. Likewise, if weight maintenance is an issue for you, be sure to discuss it with your primary healthcare provider. He/she may recommend a consultation with a Registered Dietitian (RD) to help you with diet and meal planning.

### How many times should I eat per day?

Based on advice for the general population, eating 3 to 4 times per day is reasonable. Eating less frequently can cause you to feel very hungry, which can lead to less than optimal food choices, more rapid eating, and possible choking on food. Eat enough to be comfortably full, not stuffed. Persons with HD who have higher calorie requirements may need to eat more often.

### How many calories do I need?

Calorie needs are individual, and depend on your age, gender, size, muscle mass and activity level. For a ballpark estimate of a person’s calorie needs, take their weight in kilograms (pounds divided by 2.2) and multiply by 28. That is the *low end* of their calorie range. Next multiply weight in kilograms by 40. That is an approximate *upper limit* of calorie needs. Now you have a range of calories from kgx28 to kgx40. Very few people are able to eat exactly the same amount of calorie every day. Aim to stay within the estimated range. For more specific calorie targets, consultation with a Registered Dietitian is suggested.

### How do I get there?

There are many “calorie counter” books, web-links and smart phone apps to help you keep track of calories. Choose one that you are comfortable with and track your intake.



## Tips for Increasing Calorie Intake

Here are some nutrient-dense foods to try:

- Avocados
- Smooth (not chunky) nut butters
- Sesame tahini
- Rich soups like clam chowder or lobster bisque
- Baked mashed sweet potatoes or winter squash
- Hot cereals mixed with pureed fruit
- “Better Butter” added to foods for extra calories: ½ soft butter mixed with ½ flaxseed, walnut, or olive oil

## Nutrient – Drug Interactions

- MAOIs – Avoid dietary sources of tyramine (i.e. aged cheeses, liverwurst, red wine, very ripe fruits)
- SSRIs – May cause weight loss or gain, alterations in appetite, increased or decreased glucose levels
- Anti-Anxiety drugs – May cause changes in appetite and weight
- Anti-Psychotic drugs – May also cause changes in appetite and weight.

## Mid-Stage HD

### Nutritional Needs in Mid-Stage HD

In middle-stage HD, symptoms increasingly interfere with the ability to meet nutritional needs. These symptoms may arise from the motor, cognitive or emotional disorders associated with HD.

### FAQs:

#### Are food jags normal?

Food jags, a continuous preference for exclusively eating only one or a few foods for a prolonged period of time, are a common issue with HD. Depending on the type(s) of foods being included or excluded there may be concerns about nutritional adequacy of the person’s diet. Addition of a multivitamin with minerals, or a liquid nutritional supplement may be needed.

#### How important is weight? Some studies seem to suggest maintaining weight in an HD person helps the disease.

Whatever the person’s initial body weight, weight loss raises a red flag that there is a problem which should be addressed with your health care team. In spite of the fact that not everything is known about the exact etiology of HD, and its impact on metabolism, malnutrition as a co-morbidity can and should be dealt with. Weight loss is an important indicator of nutritional status, and almost always indicates inadequate intake. Some issues may be, depression, eating or swallowing problems, or a need for a g-tube for additional nutrition support.

#### Do I need to drink Ensure®?

Ensure® is one of a number of brands of liquid dietary supplement shakes, commonly used by persons who have difficulty consuming adequate calories every day. Supplemental shakes or puddings can be added to the diet in order to boost calorie and protein intake. Like many food products, people often have their preferences as to brand and flavor, although nutritional differences may only be minor. It is convenient to have pre-mixed cans available;



they have a fairly long shelf life. Most such products are lactose and gluten free, and usually also Kosher. Some persons with HD also like to use these as a base for their own calorie enhanced beverages. A home-made smoothie can also provide easy-to-drink calories, and many recipes are available.

### **Nutrient Dense Foods / Calorie Dense Foods**

According to the 2010 US Dietary Guidelines, “Nutrient-dense foods provide vitamins, minerals, and other substances that may have positive health effects, with relatively few calories. They are lean or low in solid fats, and minimize or exclude added solid fats, sugars, and refined starches, as these add calories but few essential nutrients or dietary fiber. Nutrient-dense foods also minimize or exclude added salt or other compounds high in sodium. Ideally, they are in forms that retain naturally occurring components such as dietary fiber. All vegetables, fruits, whole grains, fat-free or low-fat milk and milk products, seafood, lean meats and poultry, eggs, beans and peas (legumes), and nuts and seeds that are prepared without added solid fats, sugars, starches, and sodium are nutrient-dense.” Foods that are low in nutrient density are foods that supply calories (energy) but relatively small amounts of micronutrients, or none at all (empty calories). The concept of **nutrient density** is beneficial advice for the general US population, where overweight and obesity are substantial health issues. However, since maintaining appropriate weight can be a challenge for people with HD, calorie-density is also an important consideration. **Calorie-dense** foods have relatively high calorie content per serving. Foods that are high in both nutrients and calories (for example: avocados, eggs, or nut-butters) are recommended to help maintain adequate calorie intakes.



### **Motor Control and the Movement Disorder**

Involuntary movements (chorea) and a reduction in control over voluntary movements may make eating a challenge. Special tableware may help.

- Dishes with sides, also called “soup plates” or “pasta dishes” can make picking up food less messy. These are readily available in most department stores.
- Sports cups with a cover and straw attached can be helpful in preventing spills.
- Spoons and forks with larger handles can make picking up food easier. Rubber sleeves that slip onto the handles of your current tableware are also available. Both of these can be bought at a medical supply store or on-line.
- A heated “keep warm dish” can be used to keep food warm throughout a meal for a slow eater.

### **Unawareness**

Another common symptom of mid-stage HD is a lack of physical self-awareness. For example, the person with HD may not realize how hungry they are until the food is right in front of them. Consistent, scheduled mealtimes can help avoid rapid eating and emotional outbursts caused by unrecognized hunger.

### **Distraction and the Eating Environment**

Distractions, noises or other activity nearby may interfere with being able to concentrate on the task at hand – eating. Certain things may be more distracting than others, so some experimentation may be needed to come up with ideal arrangements for eating. Flexibility is the key here. Some suggestions for a successful mealtime include:

- Create a quiet, calm atmosphere
- Keep the focus on eating
- Keep conversations simple – no big decision making or “hot topics”
- Allow extra time to eat the meal
- Turn off the TV, radio, or other devices
- Have enough light
- Have comfortable and supportive seating

## Chewing, Swallowing and Choking

Problems with choking and chewing may also arise at this stage. Tips for coping include:

- Try six to eight smaller meals a day instead of the usual three or four. Frequent feeding can stave off the highest level of hunger and diminish the eating urgency that can cause choking. Another approach is to offer frequent snacks or liquid supplements between meals.
- Pay attention to which foods and/or beverages seem to cause coughing or choking. Identifying problem foods can save further trouble later on. Dry, flaky or crumbly foods are often the culprits.
- Cut meat and vegetables into small pieces, and take small amounts in each bite.
- Avoid mixing liquids and solid pieces in the same bite (i.e. chunky soups).
- Choose soft, moist foods. Try mashed potatoes, noodles, soups, casseroles, yogurt, puddings, gelatins, soft cheese, ice cream, milk shakes, creamed cereals, or macaroni and cheese.
- Add moisture to foods with gravies, cream sauces, salad dressings, broth, sour cream, mayonnaise, or butter.
- Chew food thoroughly.
- Sip liquids with a straw.
- Stay away from very acidic or spicy foods that may burn the throat if irritation is a problem. (These include: citrus fruits and their juices, tomatoes and tomato juice, and spices like red pepper, black pepper, chili powder and hot curry powder)
- Add a thickener (such as ThickIt® or Thicken-Up®) or cornstarch to liquids to make them easier to swallow.

### Why are some people with HD told to drink only thickened liquids?

Coughing and choking, having beverages “go down the wrong way,” is a common problem for persons with HD. This is most likely to happen with thin liquids, like water, because these are the most difficult for the throat muscles to control during swallowing. An evaluation by a Speech-Language Pathologist can determine why this is happening, and recommend the consistency of liquids that would be best to address this problem.

Thickened liquids are an important part of a diet for safer swallowing. Some descriptive terms for thickened liquid consistencies that you may hear include: “nectar thick,” “honey thick,” and “spoon thick.”

Nectar thick liquids have slightly more body than thin liquids, but still can pour easily. Some examples include tomato juice, pear nectar, peach nectar, apricot nectar, and some bottled fruit smoothies.

Honey thick liquids are liquids that pour slowly, like honey or molasses. When you tip a spoonful of a honey thick liquid, the liquid slowly drizzles off. An example would be a very thick milkshake.

Spoon thick liquids you can eat with a spoon, like pudding or custard. If you tip a spoonful of a spoon thick liquid, it will come off the spoon very slowly or not at all. Spoon thick liquids are too thick to drink with a straw.

### If I need thickened liquids, how can I get them?

Many grocery or drug stores sell brick packs of pre-thickened liquids that are labeled as to the specific consistency of the product. They look very much like a typical juice box and are handy to grab when you’ll be away from home. Some examples of pre-thickened liquids available in this form are milk, lemonade, fruit juices, and even thickened water. If you are unable to find these at a store near you, they can also be purchased on-line and delivered to your home.

You may prefer to create your own customized thickened beverages. If so, there are products that you can add to any liquid you choose to achieve the desired consistency. Some examples are “Thicken Up”® and “Thick It.”®



These are widely available in drug stores or can also be purchased on-line. Be sure to follow the package directions for best results.

You may also want to try some other tactics for thickening liquids. For example, for shakes or smoothies you may want to add pureed fruit instead of juice, or a mashed banana or avocado. Adding powdered banana is also an option. Be sure to blend it thoroughly for the best consistency and taste.

For hot liquids such as soup, try instant mashed potatoes or instant baby rice cereal, stirred in while the soup is heating. Cornstarch or flour can also be used to thicken hot liquids, but should not be added directly because this can cause lumps. Dissolve the flour or cornstarch in a little cold water first, or stir it into melted butter or oil (what the French call a roux) before adding it to your product.

## **The HD Kitchen – Some Handy Tools**

Having the right equipment may make preparing and serving meals for the person with HD much easier. Some suggestions are listed below.

- A blender and/or food processor can be very helpful in preparing shakes, soups and sauces, and for pureeing favorite foods when a very soft consistency is needed. If possible, get one that has more than one container, to allow for preparation of multiple foods quickly.
- A juicer can be useful for a person who cannot chew fruits and vegetables well. If swallowing is a problem, try adding thickening powder or mix the juice into a thick shake or soup for a nutritional boost.
- A pastry cutter or potato masher can serve many purposes in mixing and mashing foods. These are also helpful for adding “extras” like butter or sour cream to a dish.

## **FAQs:**

### **How do you handle food away from home?**

Food away from home is an important part of many social events. For a person with HD, something that brings people enjoyment can be a source of frustration instead. In many instances, planning ahead can make this situation easier.

When dining away from home, don't forget to pack any supplements or snacks such as shakes or pudding. Thickened beverages that are available in brick packs are easy to take along too. If waiting awhile before eating is difficult for a super-eager eater, having a snack before leaving home can dial down the urgency, and make for a more enjoyable experience.

Many restaurants have menus posted on-line, so you can preview potential dining destinations in advance. Also note which of your favorites have take-out or delivery available, to have on hand for the cook's day off.

Don't forget the list of appetizers. Finger foods may often be the easiest option. Most establishments will serve 2 (or more) appetizers instead of an entrée, and bring them when everyone else's meal arrives. Call ahead and speak to a manager if you have any questions about possible substitutions, special seating arrangements, wheelchair accessibility, or any other needs.

Dining during “off peak” times will mean a less crowded restaurant, and therefore less distraction. You can also ask the wait staff to remove any unneeded silverware or glasses to simplify the place settings, and take away some table decorations, so that there are fewer things in the way. If eating an entire meal will be too time consuming or stressful, consider joining your friends for dessert instead.

It may be beneficial to cultivate a relationship with a local restaurant that can be “your place.” Restaurants value their steady customers, and are likely to be more willing to accommodate one of their “regulars.” Even persons with more advanced HD can take advantage of this kind of relationship, especially if it has been long-standing.



When going to a friend's house for a social occasion, like anyone else with special dietary needs, letting your hosts know what they can do for you can make an event go more smoothly. Remember that it is ok to say "no thanks" if you are offered a food that could be troublesome for you. You may also offer to bring an HD-friendly dish to share. Pick something yummy and make enough for everyone to have a taste. (See recipes at <http://www.hdny.org/recipes.html> for some ideas.)

### **Professionals who can help**

Who will I need to see in clinic? What do they do? How often do I need to come?

[http://www.movementdisorders.org/health\\_professionals/](http://www.movementdisorders.org/health_professionals/)

There is an excellent page about non-physician professionals and what they do on the Movement Disorders Society website.

### **Will my insurance cover:**

- a. Visit with the Registered Dietician?
- b. Supplements?
- c. Enteral feeding (nutrition directly into the stomach via a tube)?
- d. Thickener?
- e. Special foods?

Insurance companies vary as to what they do or do not cover, so this is something to check with your carrier. Unfortunately, coverage for nutrition care and specialized products is usually poor or non-existent.

## **Nutritional Needs in Late-Stage HD**

### **Difficulty Meeting Nutritional Needs**

There comes a time when a person with HD should be fed, rather than continuing to try to feed him/herself. Eating begins to require so much energy and concentration that the person with HD becomes tired and frustrated before consuming adequate amounts of food. Significant weight loss or difficulty using utensils or handling food are other reasons to limit or discontinue self-feeding.

The transition to assisted eating at mealtimes does not have to be an "all or nothing" decision; they may be able to continue eating some things unassisted, while being fed when necessary. In the right frame of mind and setting (properly positioned in a calm, quiet room, and assisted by a known caregiver), the person with HD may be able to eat more than one might expect, and perhaps be fed by mouth longer into the illness.

### **FAQs:**

#### **When is a feeding tube needed?**

Feeding tubes are usually recommended when a person is no longer able to take in an adequate amount of nutrients by mouth to maintain their weight at a healthy level. They are sometimes used to provide hydration if drinking liquids has become difficult.



## When do I need to decide about whether or not I want one?

Ideally this should be decided while the person is still able to consider the pros and cons for themselves, and well in advance of needing to have one.

## Does it hurt?

From people who have had g-tubes placed, the answer is “a little,” and “kind of like getting your ears pierced” when it is first inserted. After insertion the site may be tender for a few days. Normal use of a feeding tube is not painful.

## Can I eat when I have it?

Anything that you can eat before you get a feeding tube you can eat after you get a feeding tube. Having liquids and formula feedings via tube can take off some of the pressure to eat enough to meet your calorie needs. This can allow you to enjoy the foods that you do eat, and have less stress about it.

## About Feeding Tubes and Feeding Regimens

1. A naso-gastric tube (NGT) is threaded through a nostril, down the throat and into the stomach. This type of feeding is usually temporary, and does not require surgery to be put in. One disadvantage is that it can be pulled out accidentally. A naso-gastric tube can also interfere with swallowing, which can be a disadvantage if the person is able to eat anything.
2. A peri-epigastric tube (PEG), also called a gastrostomy tube (G-tube), is a tube that is implanted through the abdomen into the stomach. It functions in essentially the same way as an NGT. Formula flows through the tube into the stomach. It is usually for longer-term use. Since it does not pass down the throat, the patient can receive both tube feedings and an oral diet. It is often beneficial to continue oral feeding for as long as possible, even if the quantities given are not nutritionally significant, as this can provide significant psychological benefits. Another type of tube in this category is called a “button” tube. It is a very short tube attached to the stomach with a longer “snap on” tube for use during feedings. When the tube is not in use, a plastic cap covers the opening. These can be useful for people who do not receive a 24-hour feeding.
3. A jejunostomy tube (J-tube) is implanted below the stomach, directly into the small intestine. It functions similarly to tubes leading into the stomach, but with several differences. The advantage of a J-tube is that it reduces the risk that formula will come back up. This is called “aspiration”, and for someone at high risk of aspiration, the J-tube might be the preferred option. Increased probability of diarrhea, and increased probability of the very narrow tube getting clogged are some potential disadvantages.

With all types of tubes, feedings are either “continuous” or “bolus” servings. As the name suggests, continuous feedings run down the feeding tube at a fixed rate throughout the day and/or night. Tolerance problems are usually minimized when feedings are given this way. A very weak or debilitated person may need to be fed continuously in order to tolerate enough intake to meet his/her needs. Persons with HD who are at risk for regurgitation because of limited stomach capacity need continuous feeding. Most people who are just starting out on tube feedings are given continuous feedings, then gradually changed over to bolus feedings.

A “bolus feeding” is a set amount of formula run down the feeding tube at specific times during the day. Bolus feedings are essentially the equivalent of a meal consisting of formula. These usually, but not always, correspond to breakfast, lunch and dinner times. Some regimens include one or more bolus “snacks” as well, for a total of four to six feedings per day. Bolus feedings are usually more convenient for caregivers, since feedings are administered only at specific times, and larger amounts are given at each sitting.



## Some additional Guidelines for Tube Feedings:

1. Position the person so that they are sitting up, or at least so that the upper-body is above the level of the stomach.
2. Practice good sanitation. Wash your hands before handling the feeding equipment. Wash feeding bags with water. Do not use soap, as it will stick to the inside of the bag and get into the formula. This can cause diarrhea.
3. Feedings should be given at room temperature to minimize the risk of cramping and/or diarrhea. Open cans of formula should be kept in the refrigerator, and discarded if they are not used in 24 hours. Refrigerated cans should be taken out 15-20 minutes before a feeding, and allowed to warm up to room temperature.
4. Always flush the feeding tube with water after a feeding. This will help to prevent the patient from getting dehydrated. It will also prevent the tube from getting clogged.
5. If medications are to be given through the tube, use liquid forms if available. If not, always be sure that the pills are finely crushed. Flush the tube with water to wash them down.
6. If the tube seems to be clogged and a small flush of water isn't effective, a flush of about 100mls of cola may do the trick. You can also try dissolving about ¼ teaspoon of meat tenderizer in a teaspoon of water and placing it into the tube. Wait five minutes before flushing again. If none of these strategies work, contact your healthcare provider(s) for assistance.



## Resources

Nutrition and Huntington's Disease

<http://hdsa.org/wp-content/uploads/2015/04/Nutrition-and-HD.pdf>

Webinar- HD & Nutrition

<http://hdsa.org/living-with-hd/caregivers-corner-webinars/hd-nutrition/>

Recipe ideas <http://www.hdny.org/recipes.html>

Movement Disorder Neurologists [http://www.movementdisorders.org/health\\_professionals/](http://www.movementdisorders.org/health_professionals/)



# Chapter 8

## Driving, Safety and Transportation



# Driving, Safety and Transportation

Having a driver's license is generally seen as a sign of adulthood and independence. Giving up driving is seen as the opposite, a sign of declining abilities and the loss of independence. In HD families, when to stop driving can be a source of conflict. Having your arguments planned out and being ready to engage in more than one conversation may make this big change less difficult.

## How HD Affects Driving

Over time, the movement, cognitive and behavioral disorders caused by HD will have an impact on a person's ability to safely operate a car. The movement disorder will affect muscle control; the cognitive disorder will slow reaction time and affect the ability to correctly perform actions in the right sequence; and changes in behavior could trigger a "road rage" scenario of impulsive or explosive behavior behind the wheel. Caregivers are on the front line when it comes to observing changes to a person's driving abilities and in planning for the difficult conversations about reducing the amount of driving or giving it up altogether.

## Assessing the Changes

You may be noticing changes in your person with HD's driving abilities. Perhaps they are making poor choices in traffic when changing lanes or making turns. Maybe you have discovered dents and scrapes on the car or damage to the garage. You may have seen your person with HD become overly upset about a minor traffic problem. The initial question is whether these occurrences are signs of advancing HD or perhaps ordinary reactions to stress.

Take note of what you see and experience. When the time comes to talk to your person with HD, it will help to have a record to bolster your argument against continuing to drive. Documenting changes to driving abilities can be done with paper and pencil or you can use online tools to objectively assess your person with HD's driving ability, such as the seminar created by the AARP and the MIT AgeLab ([http://www.aarp.org/home-garden/transportation/we\\_need\\_to\\_talk/](http://www.aarp.org/home-garden/transportation/we_need_to_talk/)). The University of Michigan has also developed an online driving assessment program called SAFER Driving: The Enhanced Driving Decisions Workbook (<http://um-saferdriving.org/>) that is completed by your person with HD alone or by the two of you together, going step by step through the assessment process. These are only suggestions. Your own internet research may lead to other assessment resources.

## Preparing for the Conversation

Gather your allies before starting the conversation. If other family members are involved in the care of your person with HD, talk to them early on and try to get them to agree that your person with HD needs to reduce their driving or give it up altogether. If possible, you want the family to present a united front on the matter.

Before you begin any conversation with your person with HD about reducing or giving up driving, take some time to think about where and when your person with HD drives. "How am I going to go to work, the store, to church?" are usually the first questions asked by a person who is being asked to stop driving. Your first thought may be that you or family members will drive them wherever they need to go. But also look for transportation options that would give them some independence. What is the public transportation situation where they live? Are there taxi services? Does your local government run an on-call van service for the elderly or disabled? Unless symptoms make it unsafe for your person with HD to travel around town alone, try to give them as much independence as possible for as long as possible.



You may want to talk to your person with HD's doctor privately about your concerns before starting the conversation. Your person with HD may see a physician or neurologist as a neutral authority, in a way that a family member never could be. Some doctors may be willing to lead a conversation or family meeting on the issue, but understand that doctors are not trained for these conversations in medical school and many doctors report being uncomfortable getting in the middle of what they see as a family dispute. Consider asking your pastor or faith leader to speak with your person with HD or lead a family meeting.

Below are questions that the doctor or faith leader may find useful for starting the conversation with your person with HD.

### **Community Mobility**

- How much do you drive?
- Do you usually have passengers?
- Do you have any problems when you drive? (e.g., day and night vision, ease of operating steering wheel and foot pedals, confusion, and delayed reaction to traffic signs and situation).
- Do you ever get lost while driving?
- Have you gotten any tickets in the past 2 years?
- Have you had any near-misses or crashes in the past 2 years?
- Has anyone (family or friends) expressed concern about your driving ability?

### **Maintaining Driving**

- Do you get regular eye exams with a vision specialist?
- Have you checked with your doctor or pharmacist about the effects of your medications on driving ability (watch for interaction with alcohol)?

### **In-vehicle**

- Are you able to enter and exit the vehicle safely; do you use any mobility aids like a cane or walker?
- Can you sit correctly in the vehicle?
  - Upright and straight in the seat
  - Distance between chest and steering wheel 10–12"
  - Line of sight above steering wheel minimum of 3"
- Are you able to access and use safety belt at all times (across body, not under driver's arm or behind driver's back)
- Do you have adequate neck mobility for checking blind spots over right and left shoulder?

### **Getting There**

- To what extent are family or friends able or willing to provide rides?
- Are there public transportation or taxi options available to you?
- Are there things you can "trade" for a ride (making dinner, taking the driver to lunch, paying for gas)?



## Managing Emotions

Giving up one of the privileges of an independent adulthood is hard. There may be disagreements and angry words. There are a lot of emotions and memories tied up with the right to drive.

There is no right time to bring up the subject, but a calm and private setting can make a hard topic easier to talk about. If there has been an accident or near miss, try to wait until tempers have cooled before starting the conversation.

When you begin your talk about reducing or ending time behind the wheel, let the person with HD know that you understand how hard this topic is to discuss. Emphasize how their continued driving would put other people at risk. Then let them talk. Give them plenty of time to speak about what driving means to them. Allow them to be angry or sad or frightened or whatever. If they ask you a question you can't answer, tell them you will look for an answer. If they dismiss what you say and refuse to talk about it, don't argue. Just let them know that you will continue to bring it up until there is a resolution. Then wait a day or so and ask them if they are ready to talk about it. Ask family members and friends who know what you are trying to accomplish to focus on what is positive about giving up driving (safety for the public and for your person with HD) and not to dwell on what is being given up.

## The Professional Assessment and After

Your person with HD's doctor can write a prescription for a Driving Rehabilitation Program, which is a professional driving assessment typically performed by an occupational therapist with a special certification. It may be covered by insurance. The goal of the evaluation is to help a person remain as independent as possible and it looks at more than just the driver's motor skills. It includes an assessment of the psychological and social importance of driving to the individual.

When the doctor suggests the driving assessment, ask them to emphasize that the goal is to help the person with HD drive safely for as long as possible. Keep the focus on safety. If your person with HD expresses fear that the doctor wants to take away their driver's license, reassure them that physicians do not have that type of legal authority. However, doctors who genuinely feel that a person is an unsafe driver are encouraged to report that person to their state's Department of Motor Vehicles, which can result in the person having their driver's license revoked.

After the assessment, the rehabilitation specialist will report if there are things that can be put in place to help the person continue driving, such as wide angle side view mirrors, or if it is time for the person to stop driving entirely. The rehabilitation specialist who performed the assessment can help identify transportation options other than driving for your person with HD. Consider asking friends, family and coworkers if they can take turns providing rides. This is a simple way that people can help out and most people are glad to help out once in a while. Your person with HD's medical social worker may also be able to identify transportation resources in your area and other HD families may have suggestions based on their experiences.

## When the Conversation Doesn't Go Well

One symptom of HD is an unawareness of one's own symptoms. This is different than denial and your person with HD may honestly not see that their abilities have changed. They may refuse to accept that they are no longer a safe driver. They may get angry and storm off when they are asked to reduce their driving or stop it altogether. Wait until another time and try again and have others try, too.

However, if you, their friends, their doctor, and your clergy cannot get the person with HD to stop driving, you may need to take drastic action. It may be necessary to take away the car keys, sell or disable the car, or enlist the local police to explain the importance of safe driving and the legal implications of unsafe driving. When the turmoil subsides and the person finally agrees to stop driving, be sure to tell them that they are doing the right thing and how brave they are for doing it. Then tell yourself the same thing.



# Chapter 9

## Managing Juvenile Huntington's Disease in the Home



# Managing Juvenile HD At Home

## Introduction

The diagnosis of juvenile onset HD (JHD) inevitably causes surprise and sadness, but it is important to remember that there is much life to be lived after the diagnosis, and that it can include joy and happiness, laughter and meaning. The focus for care should be on doing everything to make each day as good a day as possible for the person with JHD. Family members can request the JHD Handbook that provides more detail on care or download from the HDSA website [http://hdsa.org/wp-content/uploads/2015/03/Juvenile-HD-Handbook\\_2nd-Edition.pdf](http://hdsa.org/wp-content/uploads/2015/03/Juvenile-HD-Handbook_2nd-Edition.pdf). This chapter will provide some basic A to Z suggestions and strategies for managing a child at home.

## The Big Picture

Once the symptoms of JHD start, the disease will progress gradually over many years until death. There will likely be times that things seem stable, and times that the person's symptoms, or the home or family situation, seem to be changing rapidly. It is important to plan for the future, to be prepared for the changes that are coming, so as to minimize crises or unexpected situations. However, it is equally important to focus on optimizing each day, knowing that the person with HD will never be less impacted by the disease than he or she is today.

There are two different kinds of JHD families—ones in which there is also an affected parent, and ones in which there is not (for instance, if the child with HD was adopted or for other reasons is unaware of or has no contact with the parent with HD). The stresses on a family that includes both a parent with HD and a child with HD are quite different from the stresses on a family in which the parents are free of HD. In addition, the care needs and the services available, to a person with JHD at age 5, 10, 15, 20, and 25, may be quite different. In all cases, the parent or family, hoping to care for their loved one at home, should be willing to accept any help that is available, at any point along the way. HD is a family disease, and it takes a community to assist in the family's needs.

## Stages of JHD

The scale below is an adaptation of the adult HD scale. This was published in the *HDSA Juvenile HD Handbook* and is copied below. This scale can give a family member or care provider a rough idea of where a child is in the course of the disease, but unfortunately, it cannot determine how many years the child will spend in each stage.

### The stage of HD is determined by adding the points, as shown:

|         |              |
|---------|--------------|
| Stage 1 | 11-13 points |
| Stage 2 | 7-10 points  |
| Stage 3 | 3-6 points   |
| Stage 4 | 1-2 points   |
| Stage 5 | 0 points     |

### A Functional Scale for Assessing Juvenile-Onset HD

#### A. School attendance

- 3—attends school, no special assistance needed
- 2—attends school, some regular classes, some special or modified classes
- 1—attends school, few or no regular classes
- 0—unable to attend school or work program





### **B. Academic/developmental performance**

3—reading/writing/math skills appropriate to age

2—mild decrease in academic performance but still able to take a test or to write

1—unable to write legibly but able to communicate orally

0—unable to read/write/communicate orally

### **C. Chores**

2—able to assist in age-appropriate manner with household chores

1—occasionally assists with chores

0—unable to participate in household chores

### **D. Activities of daily living**

3—performs self-cares in an age-appropriate manner

2—requires some assistance for bathing, dressing, grooming, or feeding

1—assists others who bathe, dress, or feed him/her

0—unable to assist in self-cares

### **E. Lives**

2—at home with only family assistance

1—at home/group home/foster care with assistance from non-family members

0—living in a long-term care facility

## **Community Services**

Please see the chapter on Social Services in this Guide.

## **Symptoms and Management Strategies**

In the next sections, we provide an “A to Z” list of symptoms and problems that might occur during the course of JHD. The symptoms are not given in order of how frequent or how severe they can be. Note that the kinds and severity of symptoms are very different from one person to the next, and can change over time.

### **Aggression/Agitation**

Irritability is very common in children with HD, and can be accompanied by agitation and aggressive behavior, particularly (but not exclusively) in adolescents and in males. Whenever there is aggression, the parent should think first of safety. Remove anyone who is in danger to safety, protect yourself, and call 911. If there is no significant threat to safety, then it is often best to remove the person, or everyone else, from whatever situation is precipitating the behavior. Aggression in JHD often comes in brief bursts, and subsides readily after the situation calms.

At a quiet moment, think about what precipitated the aggressive behavior. Often it is a particular event, or person, or situation that triggers the behavior, and if that event, person, or situation can be altered slightly or avoided, then the behavior may be less likely to occur in the future. It is also helpful, during a quiet moment, to decide on a plan of action for the next episode of aggression, so that both the child and the rest of the family know in advance what will happen. A relatively calm and rehearsed approach to an episode of aggression may, all by itself, reduce the child’s agitation.



Children's likes and dislikes change as they grow, and that is just as true for children with JHD. Parents frequently scratch their heads at children who scream and throw their broccoli one year, and then decide the next that it is their favorite vegetable. Be prepared for changes as the child grows up.

Talk to the doctor right away if agitation or aggression is persistent or reaches a point endangering anyone's safety. Occasionally children with JHD have very serious and challenging behaviors, which may require medications or even inpatient hospitalization if the behavior endangers the child or others. Referral to a child psychologist or child psychiatrist can be helpful in difficult situations.

## Alcohol and Drug Abuse

Removing alcohol and drugs from the home is a good first step, particularly if there is any history of alcohol or drug abuse on either side of the family. Clear family policies and expectations are important. Alcohol is particularly bad for children with JHD, who already have a disease that is causing damage to the brain. Stimulant drugs such as cocaine, methamphetamine, or ecstasy will only increase the tendency to irritable or aggressive behavior. But if a child with JHD begins using drugs or alcohol, the family, school, medical team, or legal authorities should intervene with enforcement of consequences. Finding other opportunities that keep the child busy, directing the child to a different group of friends, and spending more time together as a family can help some children. Talk to the doctor if you suspect there is a problem.

## Anxiety

Anxiety is particularly common in the early stages of JHD, when the child starts to be aware of "feeling different" or having trouble learning new things or doing things that used to be easy to do. Anxiety can come out as negativism, irritability, or depression, as the child, unable to say "I can't ride a bike anymore without feeling like I might fall", says "I don't want to go bicycling with my friend today," or gets angry or comes home sad from a visit with a friend or a day at school.

School professionals see children with their peers for many hours each day, and can be a critical source of input about a child's anxiety. Does it occur at test time? When the child has to use the bathroom between classes? Before gym, or lunch, or free period?

If there is an obvious pattern to the anxiety, then changing the environment or the situation can be helpful. Watching for signs of depression is important, as depression often accompanies or underlies anxiety. The psychologist can help the child to learn non-pharmacologic ways to address anxiety, and in more severe cases, a psychiatrist can help with appropriate medications.

## Apathy

On the opposite end of the spectrum from aggression is apathy or indifference. As JHD progresses, it will be increasingly difficult for the child to remember or get organized in time to go to school, church, or to other family events, even ones that interest him. Parents and other adults will have to keep track of time for the child, and create systems or schedules or reminders to help the child get to activities on time and with the correct equipment and clothing. A well-rehearsed pattern of activities can be helpful; it may be hard for the child to learn new things or new schedules, and a familiar routine can be reassuring.

It is important to separate apathy from inactivity due to severe depression, and some children may have both. Enlist help from other adults who see the child—teachers, counselors, and others—to help sort this out. Sometimes doctors will treat a person with severe apathy with an antidepressant, even if the person lacks other features of depression, just to ensure they aren't missing something.



## **Appetite Issues**

People with HD, both adults and children, usually have a good appetite and eat well. A persistent change in appetite should prompt a visit to the doctor to ensure that there isn't some other health problem, a psychosocial issue, or a medication side effect causing the change.

It will be up to the parents and school to ensure that the child eats appropriate and healthy foods. Children and adolescents with or without HD don't always favor nutritious foods. A child who is losing weight due to the involuntary movements associated with JHD may need to take nutritional supplements; if these are not available or are too expensive, look for milk products (ice cream, yogurt, cheese, sour cream) or nut products (with caution, as chunky foods such as peanuts are easy to choke on), as these contain protein as well as extra calories.

Please read the chapter on Nutrition.

## **Bad Behavior/Cursing**

Some children have episodes of disruptive behavior such as cursing, oppositional, or defiant behavior. These behaviors, even though they are less dangerous than agitated or aggressive behaviors, can provide a parent with challenges.

The parents have to "choose their battles" with regards to defiant behavior. The best way to get a child to cut his ridiculously long hair may be to ignore it. But if church attendance is important and the child repeatedly ignores a mother's requests and demands to get dressed to go, then perhaps the father can be put in charge of getting the child up and dressed for church. Or perhaps there can be a compromise, such as going to a later service.

Young children often respond well to "time outs" as a strategy to manage bad behavior. Older children need consistency. If one parent punishes the child for cursing, and the other one thinks it is funny, the child will get a mixed message and will quickly learn to play one parent against the other.

Family counseling can help parents learn strategies to manage bad behavior.

## **Chorea**

Young children with JHD are somewhat less likely than adults to have chorea. Adolescents or young adults may develop chorea as part of their movement disorder. "Chorea" refers to dance-like involuntary movements of the limbs, trunk, head/neck, tongue, or diaphragm. Mild chorea may look like fidgeting. Severe chorea can make it difficult for a person to sit, stand, or walk, and may contribute to speech disturbances and clumsiness of the hands.

Chorea in people with HD tends to be worse with stress. Severe limb chorea may be misinterpreted as hitting or kicking, and erratic gait due to chorea can be misinterpreted as being due to intoxication. We encourage adolescents or young adults with HD who are active in the community to carry a wallet card that briefly describes HD, in case a law enforcement situation arises, and to rehearse in advance what to say if they are stopped by a police officer. HDSA offers training tools for law enforcement that are available online for downloading. Go to <http://hdsa.org/living-with-hd/law-enforcement-education/>

HDSA also offers an HD ID bracelet that can substitute for a wallet card. Look at the HDSA bracelet on the HDSA.org website. <http://hdsa.org/shop/accessories/>

## **Communication Challenges/Speech/Dysarthria**

See the chapter on the Speech-Language Pathologist.



## **Dementia/Comprehension Issues/Confusion/Forgetfulness**

Dementia is a feature of HD which particularly affects the domain of “executive function.” Executive functions include the ability to make decisions, multi-task, initiate activities, and suppress undesirable thoughts or behaviors.

Please refer to the HDSA publication, *Understanding Behavior in HD/JHD* for more information about this.

For the younger child with JHD, dementia will lead to an ever-growing disparity between the child’s academic abilities and those of his classmates. Depending on the age of onset, there may come a time when the educational goals shift from learning academic material to learning or maintaining social and independence skills. The family should work with the school to develop an Individualized Educational Program (IEP) that is suited to the child’s cognitive abilities. Because HD is a progressive disease, last year’s goals or plans may not be appropriate this year. Though some children may show more rapid changes that necessitates significant changes in the IEP mid-year; in most instances, an annual plan will suffice with only minor mid-year adjustments.

## **Dental/Oral Hygiene Issues/Drooling**

Children with JHD should have at least the same attention to dental matters that a child without HD has, if not more. This is a disorder that leads to difficulty using the lips, tongue, and muscles of the mouth, causing difficulty with speech and with chewing and swallowing. These problems will only be made worse by years of poor oral hygiene, leading to painful gums and even loss of teeth. Children with HD may have difficulty with the act of tooth brushing, and may also resist this seemingly minor but important act of hygiene.

Parents can help the child with JHD by creating an expectation very early on and a daily routine that includes oral hygiene activities.

Drooling can be a concern for some children with HD. Reminders to keep the mouth closed or to swallow may help. Medications are sometimes used to “dry up the saliva,” but they can have side effects such as dry mouth, constipation, and cognitive slowing, and so have to be used carefully.

## **Depression**

Depression is common in HD and should be recognized and treated aggressively. Symptoms of depression include a loss of interest in activities that a person used to enjoy, a change in appetite (in either direction), a change in sleep habits (in either direction), tearfulness/crying, negativity, and in severe cases, changes in cognitive function, suicidality, or psychosis (paranoid or delusional thinking).

Depression is readily treatable in most cases with counseling or antidepressant medications or the combination of both so parents should not hesitate to let the doctor know if they suspect it in their child.

## **Dying and Death/Hospice**

Death from HD generally comes after a number of years — 10-20 years, perhaps more — and only when a person has reached the late stage of the disease. Warning signs that a person may be reaching the end of life include: wheelchair- or bed-bound, minimally able or unable to communicate, losing weight despite aggressive efforts on the part of family and caregivers, recurrent infections or hospitalizations, and an unusual increase (for that person) in agitation.

If a family’s goals for the child are that the child be comfortable, in a familiar place, free of pain and surrounded by family, then the family might want to invite a hospice team to participate in the child’s care, as these are the general goals of hospice. It is helpful to understand when death is coming, to allow for a comfortable, dignified end, and a celebration of the life that has preceded it.



## **Dysphagia/Eating**

See the Nutrition and Speech-Language Pathologist chapters.

It is important for the young adult with JHD, or the parents on behalf of a younger child, to determine whether they would want placement of a feeding tube at a time of increased weight loss or increased swallowing difficulties. A feeding tube can reverse weight loss, and provides a reliable way for calories, liquids, and medications to be given. A feeding tube is generally inserted by a radiologist or gastroenterologist (GI specialist) as a same-day procedure, with the patient sedated. A feeding tube can easily be managed by an adult in the home setting, with minimal instruction from a nurse. Families can discuss the pros and cons of a feeding tube with their doctor.

## **Hallucinations/Paranoia/Suspiciousness**

Hallucinations, paranoia, psychosis, and delusional thinking are uncommon in both adult and juvenile-onset HD, but when they occur they are quite disturbing to the family, friends, and community. The acute onset of hallucinations or psychosis is an emergency, as it could be caused by a dangerous level of drug intoxication, or could occur in the setting of a severe metabolic problem. If the problem is more chronic, and other causes have been ruled out, then the psychosis is probably related to JHD. Primary care doctors and neurologists are generally uncomfortable managing this dramatic psychiatric symptom, so most children with hallucinations or psychosis are referred to a psychiatrist for evaluation and management.

## **Incontinence**

Control of the bowel and bladder are generally not major issues in the early stages of JHD, unless the child is very young and has never fully gained control of these activities in the first place. In the middle stages of the disease, affected individuals tend to become less attentive to their hygiene, and so may not bathe or brush their teeth, comb their hair, or attend to toilet hygiene as well as they should. But loss of bowel and bladder control do not usually occur until late in the disease, around the time that mobility is becoming increasingly difficult.

## **Independence**

The primary goal of the school and health professionals is to allow the child to develop to his or her fullest potential despite the presence of JHD, and to permit the child to have as much independence as is safe, for as long as possible. For the family dealing with JHD, this process is especially challenging, because the child may be growing and developing in some ways, while remaining static or declining in others. A plan of care that works well one year may not work the next, as both the child and the disease change. Parents also have to be available or have made plans for that time in the future, when the child will be less independent than he or she is currently, so that greater assistance can be added as smoothly as possible.

## **Obsessive/Compulsive/Perseverative Behavior/Fixations**

It is common for persons with HD to “get stuck on an idea” and have difficulty switching to another topic. Young children normally tend to be this way, so the behavior may not be too unfamiliar to the parent. Obsession implies that the person derives some pleasure from the thought, or that it reduces some kind of discomfort (a person who has an obsession about cleanliness, for instance, might only be happy using new freshly cleaned towels every day; while a young woman obsessed with a teen idol may be happy to see his face on the (many) posters on her wall). Perseveration, on the other hand, is just repetitive behavior, often related to cognitive changes (a 3 year old who asks “are we there yet” every few minutes simply lacks an understanding of time and geography; a 15 year old with JHD asking the same question on the way to a familiar destination has probably lost that understanding).



Perseveration can often be managed by distraction or redirection. Obsession, particularly when it is accompanied by compulsive behaviors (the person obsessed with cleanliness who washes their hands multiple times before leaving the house, for instance), may respond to cognitive-behavioral therapy, but often requires medication to reduce the intensity of the urges. A psychiatrist commonly manages obsessive-compulsive behaviors.

Families have to find a middle ground between allowing obsessive-compulsive behaviors to rule the household, and coming to blows over how many times a child washed their hands before leaving the house. Family counseling may be necessary to help the family find that middle ground.

## **Pain**

HD, by itself, does not cause pain. An occasional child who has severe dystonia (muscle stiffness) may have muscle pain. And obviously, if there are cuts or bruises or broken bones due to falls or near-falls, this can cause discomfort. But HD does not cause headache, stomachache, painful breathing, urination, or passage of the bowels. The new development of pain should be evaluated and treated as it would be for a child who does not have HD.

## **Rigidity/Dystonia**

Muscle stiffness is more commonly seen in JHD than in adult HD for reasons that have not yet been explained. Doctors may use the words spasticity, rigidity, or dystonia to describe different kinds of stiffness. Stiffness in the legs can cause a jerky, straight-legged gait, or toe-walking. Stiffness in the trunk or the arms can cause unusual postures, or slowness of movement. Stiffness can also affect the neck and muscles of the mouth and throat and lead to changes in speech (dysarthria), chewing and swallowing (dysphagia).

Stretching, then application of heat and massage, can all help temporarily to relieve tight muscles, and should be part of the fitness regimen for a person with HD. Doctors sometimes use antispasticity medications depending on the situation and severity of the symptoms.

## **Seizures/Epilepsy/Myoclonus**

Epileptic seizures almost never occur in adults with HD but occur in about 25% of children with JHD. The types of seizures are variable, ranging from very brief staring spells or single jerks of a limb or the trunk (myoclonic seizures), to generalized tonic-clonic seizures (in which the child is unconscious and shaking repetitively for up to 3 minutes, and often dazed or confused afterwards). Any child who has a seizure for the first time, or who is suspected to have had a seizure, should be evaluated with brain imaging (an MRI, or magnetic resonance imaging) and an EEG (electroencephalogram). Blood and urine tests done in the emergency room at the time of the seizure can rule out a low blood sugar, drug intoxication, infection, or other metabolic causes.

No medication comes with a guarantee that it will prevent seizures, and all antiepileptic medications can have side effects. Parents need to work with the treating neurologist to find a drug that has maximum benefits with the least side effects. Stress can increase the likelihood of a seizure, whether it is stress due to an illness such as the flu, an emotional stress, lack of sleep, or a low blood sugar.

## **Sexuality**

Adolescence is a difficult time, even without JHD, as a young person develops adult sexual features and feelings, with no experience in how to use and control them. If JHD is developing at the same time, it can make the advent of sexuality even more confusing and challenging. Teenagers with JHD should be considered sexually vulnerable—either





to the advances of others or to poor control of their own sexuality. Parents should help their child use appropriate contraception if they are sexually active or enlist the help of the doctor or the school to ensure that they are protected.

Children who develop JHD prior to adolescence may be somewhat easier to manage with regards to sexuality, as they may not develop the intense urges that come with adolescence as they get older, but rather may remain emotionally in a preadolescent state. These children, in particular, may be vulnerable to inappropriate sexual advances of others, and may benefit from having adult supervision in the school locker room or bathroom.

Girls who develop JHD in childhood or early adolescence may have difficulty managing their own menstrual hygiene; long-acting hormone preparations that suppress menstrual periods may be helpful in this circumstance.

## Sleep Disturbance

It is often hard to determine whether sleep disturbance in a teenager is “caused by HD,” related to a complication of JHD such as depression or a medication side effect, or whether it is “just the child.” Parents should do what they can to train the child to use good sleep hygiene, which includes the following:

- Have a regular time to go to bed and wake up.
- Sleep only in the bed, and only sleep in the bed (i.e., no reading, playing computer games, eating, watching TV while in bed).
- Be active in the early evening, but then engage in quiet activities to “wind down” before bedtime.
- Avoid stimulant drugs (such as ADD or ADHD medications), coffee, or caffeine-containing sodas, at or after dinnertime.

If there is a sense that a newly-prescribed medication is causing excessive sleepiness (or inability to sleep), contact the doctor who prescribed the medication. If other features of depression (or the opposite, mania) appear to be present, contact the doctor right away.

If, despite all these things, the child still tends to stay awake later than they should, and then has difficulty getting up for school the next morning, know that this pattern is extremely common in children who do not have HD! Parents may have to pick their battles, perhaps even compromise with their adolescent, on an acceptable sleep and waking time. Consistency is important. If the child visits a grandparent, or alternates visits with one parent or another, the acceptable sleep and waking times should remain the same.

## Stress

Stress is all around us, particularly for children, whose bodies are growing and changing and who are struggling to break free of their parents, to develop friendships, to discover their talents and weaknesses, and to succeed. School is a competitive environment, and often so is home. A chaotic home environment can include one in which there is a parent and a child with HD, possibly other children, an exhausted spouse/parent who may be working two jobs while managing a spouse and child with HD, the other children, and all the usual household chores. Even without an affected adult in the home, there can be stresses within adoptive or foster families.

Children deal with stress in many different ways. Some become irritable or aggressive, while others withdraw to their own private world. Some avoid their home and/or skip school and hang out with their friends who are having similar issues. A family with HD should be assumed to be a family under stress, and the child needs to have an outlet outside the home (such as a school counselor or a Big Brother/Sister). Children recognize stress in their parents and families; not talking about it teaches the child that there are family secrets, that this family does not acknowledge stress, and forces the child to turn elsewhere to seek relief.



Family members will need support, too. In some communities, there are HD support groups, or HD lay organization chapters, such as HDSA, that can help parents or families. In other communities, parents may have to join together with parents of children with other sorts of neurological diseases or handicaps to share mutual experiences and challenges.

Healthy children in HD families can turn to the HDSA National Youth Alliance <http://hdsa.org/about-hdsa/national-youth-alliance/> or other organizations such as the HD Youth Organization ([www.hdyo.org](http://www.hdyo.org)) for information and support presented in a kid-friendly manner.

## **Suicidality**

The risk of suicidal thoughts and attempts are both increased in persons with HD compared to the general population. Risk factors for suicide include depression, drug or alcohol use, impulsiveness, family history of suicide, previous suicide attempt, and a sense of isolation or hopelessness. While some of these risk factors cannot be changed, some of them can. Another important risk factor is access to the means, such as firearms or a stockpile of medications.

The family can help reduce the risk of suicide by treating depression aggressively, asking about suicidal thoughts, removing weapons from the house, and securing car keys, medications, and alcohol where the child cannot access them. Counseling for the child and the family may be helpful to identify other solutions to the child's challenges that do not include self-inflicted death.

Suicidal behavior is an emergency; the family should call 911 and stay with the person, if it is safe to do so, until the emergency responders arrive. Children or young adults who are having persisting suicidal thoughts or a suicide attempt may need hospitalization in order to stabilize their mood and to remove them from their usual environment.

## **Walking/Falling/Balance**

The movement disorder that is part of JHD invariably affects a child's ability to walk. When the disease symptoms begin in the first decade, a stiff-legged, jerky gait may be one of the first symptoms. In the first 10 - 15 years of life, children normally gain motor skills — first walking, then running, riding a tricycle, a bicycle, and then doing more complex activities such as skateboarding or skiing. A loss of previously acquired skills in a child is an ominous sign, and may suggest the onset of JHD.

Children need to remain active as they fight JHD, but they also need to be safe, and to succeed at the activities that they are doing. A physical therapist can help to design an exercise regimen that is safe and addresses the child's neurological issues, and a personal trainer or gym instructor can adapt this using the resources of the school or fitness center. While the physical therapist often focuses on gait, balance, and large muscle movements (legs and trunk), an occupational therapist can work with the child to optimize fine motor skills (hands and fingers), as JHD does affect both areas. Children should be permitted to be as independent as possible, as long as an occasional fall without injury is not turning into daily falls requiring trips to the emergency room.

Although the goal is to keep the child walking for as long as possible, in the late stages of the disease, most people with HD are no longer able to walk on their own. If the goal is to keep the child in the home until the end, then the parents should think about handicapped accessibility—the ability to get a walker or wheelchair into the house and maneuver it in the spaces in which the child is living.

## **Things to Plan For and Do**

This section will describe briefly a variety of things to consider as a parent or family cares for a child with JHD in their home. This is not an exhaustive list but hopefully it will provide some useful suggestions and tips.



## Activities/Games/Leisure

One of the things that is forgotten very quickly when a family is stressed is the whole idea of having fun! When the day is filled with challenges and problems there seems to be little time for games or leisure activities.

Children need games and puzzles and leisure time. They learn by playing games, they relax and relieve stress and have pleasant memories of leisure activities such as trips to the zoo, the baseball game, the grandparents, or the Air and Space Museum. When designing activities for a child with JHD, it is important to ensure the activity is appropriate cognitively and physically.

The Make a Wish Foundation, and other similar organizations, can help a family design the dream of a lifetime for a child with JHD. Take that trip to Washington DC or Disney World, or meet the football player or teen idol of your dreams!

Make scrapbooks filled with pictures of family members smiling, laughing, or doing silly things. In difficult times, those pictures might just bring a smile to someone's face.

Later in the disease, as it becomes more difficult for the child to participate in activities outside the home, there will inevitably be more time available for such passive activities as TV watching and game playing. The family will have to decide at that point how or whether to restrict the child's time with these activities and provide alternatives.



## Alternative Therapies/Vitamins and Supplements

Whether it is out of frustration with regular medical care, or a desire to try everything that might be helpful, many people turn to “alternative treatments,” vitamin supplements, Eastern medicine, homeopathy, and supportive therapies such as massage and yoga. It is impossible to make a blanket statement about all of these non-physician prescribed treatments, because they range from foreign chemicals to plant or animal extracts, to therapies performed by practitioners of various types.

Several vitamins and supplements have been shown not to have an effect in HD in large clinical trials, including creatine, Coenzyme Q10, and the active ingredient in fish oil. There have not been research studies conducted of most compounds for treating HD symptoms. It is important to review whatever the child is taking with the medical doctor, as some innocuous-sounding compounds can interfere with prescribed medications and the chemicals in “natural supplements” can have both good and bad effects in the body.

If a family decides to try chiropractic, massage, yoga, homeopathy, acupuncture, or any other treatments or therapies, make sure that the practitioner is certified in the area of specialty, much as you might check on the medical doctor. If a therapy seems to help, then continue it—but if it is expensive and shows no benefits or causes problems, then stop the treatment.

## Bathroom Safety

The bathroom is the most dangerous room in the house for people who tend to fall, and the bathroom in a junior high or high school can be a frightening place for a child with JHD. Children who have trouble with toilet hygiene or managing their clothes, or who feel uncomfortable in the bathroom because of interactions with other students, should have a care plan that includes a personal care attendant to assist them.



The occupational therapist can make suggestions to the family about home safety including proper equipment and strategies to make the bathroom as safe as possible.

## Care Facilities

Sometimes, despite everyone's best intentions and commitment, it can become impossible or inappropriate to manage a child with JHD at home. It is important that no one think of this as a failure. There are several very different scenarios that might lead to placement outside the home.

Even with JHD, children grow up to become young adults, and are aware of the excitement that classmates have about growing up and moving out of the home. They may prefer to live in a group home with people their age, rather than at home with their parents. It is important to take the child's views into consideration, knowing that there may be additional financial or logistical considerations that influence the decision.

Some adolescents with JHD have severe and persistent, treatment-resistant behavioral problems. Particularly if there are other children in the house who are either directly at risk because of the behaviors, or whose needs are set aside because of the dramatic challenges of the JHD-affected child, the parents may have to make a very difficult decision. Sometimes, for the sake of the rest of the family, the decision is made to move the child with JHD out of the home to a residential school or foster home. Nobody makes a decision like this lightly, but it is important to consider the needs of the whole family, and not just the child with JHD.

Finally, if the primary caregiver has health problems, physical or emotional, particularly if they are expected to be chronic, then it may not be possible or desirable to keep a child with handicaps at home. This obviously depends on the severity of the caregiver's illness, as well as the stage of disease and typical symptoms of the child with JHD.

If the plan is to keep the child with JHD at home then, it is important to have a backup plan. What if the caregiver becomes sick? Talk to the county, school, or HDSA chapter social worker to learn what residential options exist in your area.

In some communities, respite care is a possibility. Respite care could be a professional person coming into the home for a few days or a week, or the person with JHD going to a facility temporarily, to give the person and the family a break from each other. If the experience goes well, then it makes it much easier to make a more permanent transition later.

## Celebrations/Birthdays

Birthdays and family celebrations take on even greater meaning in the presence of a neurodegenerative disease, serving as a reminder that another year has slipped away, never to return, but also as a cause for celebration as the family fights HD. So, celebrate. Take pictures! Make scrapbooks or memory books to remind yourself and your child about the joyful times together. Reviewing those scrapbooks can bring happiness to a child at a difficult time later in the disease.

## Dignity

Even a child needs dignity. A child should not have to tolerate taunting by classmates or family members. Personal care assistance should be provided privately with compassion. An exhausted caregiver should seek respite rather than snapping at the child who is slow or doesn't understand or doesn't quite do what they are supposed to do.

All adults should treat the child with JHD with courtesy and respect. In this way they will serve as role models for children who aren't sure how to respond to someone who isn't quite like them.



## Dressing and Bathing

Dressing and bathing are two of the activities of daily living that become difficult for children in the mid-stages of JHD, for different reasons in different children. Some simply forget or have difficulty organizing the complicated set of tasks that are required. For some, it is a behavioral issue. And for others, poor coordination or slowness prevents the child from completing the activities in a timely manner.

Helping the child to get dressed requires some delicacy, as a parent tries to allow the child to maintain as much independence as possible while providing enough support to get the job done. Some children just need help selecting appropriate clothes; others need an electric toothbrush to make that task a little easier. It is only late in the disease that a child would be expected to have incontinence or need total assistance with bathing and dressing.

## Driving

Getting a driver's license is a major rite of passage for 16 year olds in most towns and cities in the US. A child affected with JHD prior to that age may have difficulty passing either the written or the road test, which will show once and for all that the child is no longer keeping up with his or her peers. Parents who can see this failure coming can prepare their child for it, possibly steering the child away from driver's education in the first place, or downplaying the importance of the license.

Almost more problematic is the child who is just beginning to develop JHD at age 16, who is able to pass a driving exam and get a license, but who just a few years later may lack the insight that by then makes continued driving unsafe, and continues to drive.

An older teenager or young adult who is not going to drive needs an alternative means of transportation, preferably one that does not involve his parents.

## Emergency Pack

Unlike many childhood diseases, like asthma and diabetes, acute attacks are uncommon in JHD. However, older teenagers or young adults who are in the community independently should carry a wallet card or wear an ID bracelet that explains briefly what HD is, as the gait abnormalities in HD are sometimes interpreted as "walking like a drunk" by bystanders who call the police.

Two emergencies can occur in a child with JHD. The first is a seizure, which can occur in 25% of children with JHD. Ask the doctor what emergency medications, if any, should be available at home to treat seizures, and under what circumstances a child with a known tendency to seizures needs to come to the emergency room. The other emergency situation that could arise is a behavioral crisis, with aggressive, impulsive, suicidal, delusional, or other disruptive, dangerous, or disturbing behaviors. As-needed sedative medications might be provided for anxiety or aggression. But more commonly, the child needs to be removed from the situation that is triggering the behavior, or the help of others may be needed if the behavior is severe.

## Environmental Adjustments and Equipment

As JHD progresses, it may be more difficult for a child to go up and down stairs, or the person with JHD may stumble into coffee tables or trip on throw rugs. Children with severe chorea may need a recliner with padded sides to avoid bruising. An occupational therapist can evaluate rooms in the house and daily activities, identifying problem areas or dangerous settings, and make recommendations about major or minor changes to the environment that can reduce the risk of falls and other injuries.



## Exercise

Exercise is critical for everyone who has a movement disorder. The details are not critical; we have not determined that one kind of movement is “more effective” than another, or how many minutes of exercise are necessary to have a beneficial effect. But it makes sense that a person will be able to move and balance and walk if they have been doing those kind of movements. A physical therapist or exercise professional can help the family work out a specific exercise plan that is varied and enjoyable.

## Friends

It will likely be difficult for a child with JHD to maintain friendships over the course of the disease. As regular schoolwork becomes more difficult, the child will likely start to be grouped with new friends, often other children with various types of handicaps. Parents and school professionals will have to work carefully to balance the child’s social desires with what is appropriate, feasible, and safe. Dating may be difficult, but attending school activities with groups of friends may work well for some children.

A few children with JHD have created a strong internet presence through blogs, Facebook, or other kinds of social media. Parents need to monitor any internet activities to ensure that the child is using the internet appropriately and safely.

For some children, attendance at the HDSA national convention can be an exciting annual adventure as they see other children with JHD that they have met at previous conventions. The convention can also provide support and education for family members.

## Future

Focus on the short-term future can include the child and family being active participants in the HD community; as advocates, support group participants, research participants, or other means that fit with the family’s talents and interests.

The families that have the most hope for the future as it relates to HD are those that are engaged in the HD community in one way or another. Children who participate in research studies, give speeches about HD in their school or in HD groups, attend the HDSA national convention, organize or attend fundraisers elevate everyone around them and promote hope for the future.

Family trips and adventures can also give the child things to look forward to in the future. And if the child has somewhat greater goals, such as moving out of the house, driving, getting a job, or marrying and raising a family, parents should support the child in achieving the interim steps and goals that make might make those things possible without making false promises.

## Home Safety

Adapting the home for safety is one of the most important things that a family can do to optimize a child’s independence throughout the course of JHD. As the disease progresses, mobility, coordination, and communication are all impaired. By thinking in advance about how these changes impact the activities of daily living, and adapting the house accordingly, the family will allow the child to succeed, and be safe and comfortable in the home.

## Hospitalization

If hospitalization is necessary, because of an infection or an injury or a behavioral crisis, it is critical to work with the hospital team to let them know what challenges the family thinks will impact hospital care. Sometimes the hospital will allow a family member to stay in the room with the hospitalized child.





A hospitalization may herald a new stage in the disease when the strategies and systems that worked in the past don't work anymore. It may be necessary to confront the idea that the child may need more help, temporarily or permanently, after discharge from the hospital.

This may be a time to ask friends and family to make a trip to the hospital, pick up other children at school, prepare a meal, or come to the hospital so that a parent can get a break for sleep or a shower. This is a good time to ask for help if a parent feels overwhelmed.

## School

The school can be the family's greatest resource in the day-to-day management of JHD. It provides a supervised place where the child can go to have cognitive and physical exercise and social interactions. Public schools are required to work with families and the medical team to provide an independent educational program for children with all sorts of handicaps, and to adapt the child's curriculum and school activities to meet the child's needs for maximal independence. The school can also help the young adult child and family transition into an appropriate post-school program or living situation. Finally, the staff in the school that knows the child can provide support not only to the child, but also to the parents and caregivers. Independent educational programs for children with JHD have included modified classroom programs, modified physical education activities, a personal care attendant to assist the child in the bathroom and with transitions between classrooms, independent skills training, and coordination of staff to optimize the child's learning, pleasure, and safety.

### FAMILY NOTE

*My daughter was the homecoming queen for her school!*

## Transitions

All children develop, grow and move from young children to young adults. JHD impacts on that development, while other family issues may impact on how a family can address these transitions. It is impossible to describe a simple formula for handling transitions in HD.

In general, however, children go from being playful, loving young children to world-watching preteens, to conflicting and conflicted teenagers, to tentative young adults trying to start out on their own. Depending on when HD begins there are obvious effects on the degree of cognitive, physical, and behavioral symptoms it causes. Children whose emotional development peaks in the pre-teen stage may be very happy (but vulnerable), playful individuals who freely accept help and guidance from the adults around them. Children who have reached adolescence as HD hits may have much more tendency to anger, substance abuse, and severe antisocial behavior, and may resist help and support from family and school. Still others may reach a stage in which their main goal is to live independently and work, and just as with children who don't have HD, parents may do best to allow them to try to do this, while providing a safe haven in case the attempt fails.

The best way to make transitions smooth is to plan ahead. Whether a person is moving from an upstairs bedroom to a bedroom on the main floor to avoid falling on the stairs, switching from junior high to senior high, starting to use a wheelchair, transitioning to a group home, or needing in-home care from a professional provider, if the parents are prepared for it, present the change in a matter-of-fact way, and gently but firmly guide the child to making or "approving" the decision or transition, then things will likely go more smoothly.

And some transitions, despite best efforts, will be bumpy. Support is available from family, friends, a support group or a counselor, to get through these difficult times. An HDSA social worker can talk with you about local support groups or may know other families in a similar situation. You need not feel alone.



## RESOURCES

Locate a social worker

<http://hdsa.org/about-hdsa/locate-resources/>

The Juvenile HD Handbook [http://hdsa.org/wp-content/uploads/2015/03/Juvenile-HD-Handbook\\_2nd-Edition.pdf](http://hdsa.org/wp-content/uploads/2015/03/Juvenile-HD-Handbook_2nd-Edition.pdf)

Juvenile Huntington's Disease and the School Experience: Education and the Child Affected by Juvenile Onset HD

[http://hdsa.org/wp-content/uploads/2015/03/JuvenileHD\\_School-Experience\\_APR2015.pdf](http://hdsa.org/wp-content/uploads/2015/03/JuvenileHD_School-Experience_APR2015.pdf)

Talking with Kids About HD

<http://hdsa.org/wp-content/uploads/2015/05/TWKBrochure-FINAL-WEB.pdf>

Law Enforcement guides

<http://hdsa.org/wp-content/uploads/2015/05/HDSA-LET-Caregivers-Guide1.pdf>

<http://hdsa.org/wp-content/uploads/2015/05/HDSA-LET-Guide1.pdf>

HDSA medical alert bracelet

<https://interland3.donorperfect.net/weblink/weblink.aspx?name=E5596&id=150>

HDSA National Youth Alliance <http://hdsa.org/about-hdsa/national-youth-alliance/>

HD Youth Organization ([www.hdyo.org](http://www.hdyo.org))



# Chapter 10

## Interacting with Health Care Providers



*This chapter will assist caregivers as they interact with different healthcare systems like the doctor's office, the hospital and the mental health system to provide quality care for the person with HD while trying to manage a potentially stressful situation.*

## Interacting with Healthcare Providers

Interacting with healthcare providers (HCP) can be intimidating. However, it's important to remember that when it comes to Huntington's disease (HD) and your person with HD, YOU are the expert. Healthcare providers need you to help them understand HD and your person with HD's unique needs and preferences. The purpose of this chapter is to help you feel confident in your role as an advocate for your person with HD when interacting with healthcare providers from inpatient clinic visits to long-term care. This will help to ensure your person with HD receives the best possible care.

### Outpatient Visits

If you live near an HDSA Center of Excellence (COE) or are able to travel to one once a year, you can be confident the HCP are knowledgeable and experienced in caring for persons with HD. Even so, each person with HD is unique, and you know more about your person with HD than anyone else. Your involvement in their care is essential. If you are unable to visit a COE, the best type of HCP to treat a person with HD is a movement disorder neurologist or a psychiatrist. These professionals might not have taken care of a person with HD before so your involvement in helping to provide them with information about HD and your person with HD is important. Ask your person with HD's primary HCP for a referral to one of these specialists if you do not already have one. In some areas this might not be possible. HDSA can help refer you to a HCP that has experience with HD or can serve as a resource to help your person with HD's HCP know more about HD.

Some HCP who are not experienced in caring for people with HD might not know that HD is a treatable condition. Be prepared to help educate the HCP about HD and that there are effective treatments for symptoms. HDSA has a publication, *A Physician's Guide to the Management of Huntington's Disease*, that is available <http://hdsa.org/shop/publications/#clinical-care>. In addition, HCP might not understand that HD is not just a motor disease, but that it also affects behavior. HDSA also has a publication for HCP called *Understanding Behavior in Huntington's Disease: A Guide for Professionals*. In addition, your HCP should have access to medical journal articles about treatment for HD symptoms. Some suggested articles appear at the end of this chapter.

It is important to be engaged in your person with HD's healthcare even if he or she is still able to attend appointments independently. One day you will have to be more actively involved in making care decisions so it's important to develop a positive relationship with your person with HD's HCP early. In addition, you might know important things about your person with HD of which he or she might be less aware. It is common for people with HD to lack awareness of some of the behavioral symptoms such as losing interest in activities, irritability, forgetfulness, driving safely, or performing activities of daily living. If you are not present to tell the HCP, the HCP might not know these issues exist.

#### FAMILY NOTE

*Brandon Rogers, former president of the HDSA Iowa Chapter, whose mother died from HD complications, said, "become an expert" in your loved one's disease. Journal how days go, what interventions work or do not work. You are the voice for your loved one.... You are their advocate and you need to be there for them. Ask questions, [such as] 'Why are we trying this medication?' 'Could they benefit from physical therapy?' 'They have been really "flat" lately...is there anything we could do to help them?'"*

#### FAMILY NOTE

*An HDSA chapter member caring for her mother with HD recommends the whole family attend clinic appointments, "so if one person forgets something or is possibly in denial about a new HD symptom, the other family member is*

*there to fill in the physician.” At the same time, it is important to let your loved one speak for himself or herself as much as possible. She suggests that the family can often be helpful in ‘translating’ for the person with HD. “Do be careful to keep the HD person in your focus as they are able to understand but it may take a moment for them to respond to your question. Be patient and pause to get a response directly from them when possible. They want to feel like they matter and have a part in their health care, just as you would.”*

If you feel your person with HD might not present symptoms accurately to the HCP, consider sitting or standing behind him or her so that you can gesture to the HCP when you don't agree with what your person with HD is reporting. If you don't feel comfortable doing this, or talking about your person with HD's symptoms in front of them, request before the appointment to have time to talk privately with the HCP or arrange a follow-up phone call.

Another important reason to be involved in your person with HD's treatment is to provide feedback to the HCP on whether treatments are effective and whether there are side effects. Again, your person with HD might be less aware of these issues than you are. It's important that treatments are aimed at improving symptoms that are the most important to your person with HD and to you. Communicate with the HCP about what changes, if they were possible, would best improve quality of life.

For more information on this topic, HDSA has a guide in its Family Guide Series called “A Caregiver's Guide to Communicating with Healthcare Providers,” which is available on the website.

## **When to Seek Medical Treatment**

In addition to regular outpatient appointments, it might be necessary to seek medical treatment in more emergency situations. It might be difficult to know when something is a medical emergency. Since HD is a slowly progressive disease, it's sometimes difficult to know exactly when it's important to seek medical treatment or advice and when to try to manage on your own. In general, it is important to get an evaluation by a HCP if a change in behavior or health is sudden, or your loved one is in pain, or your safety is a concern. A change in health status could include a fever and coughing. Or, a sudden change in behavior could include confusion, disorientation, or aggression. The most common reasons for hospitalization in the later stages of HD are pneumonia (often as a result of aspiration), psychiatric reasons, dehydration and nutritional deficiencies, pressure ulcers, trauma related to accidents, and infections such as urinary tract infections and infections in the blood.

Fever and coughing could indicate pneumonia. Pneumonia can occur if swallowing becomes difficult and food and liquids can accidentally enter the lungs. If your person with HD is coughing frequently and develops a fever, he or she needs to be evaluated for pneumonia. If your person with HD is choking more often while eating (dysphagia) but is not coughing, and does not have a fever, it's important to schedule an appointment with a neurologist. There might be ways to decrease swallowing difficulties with therapy and/or diet changes.

If more frequent falling is an issue, physical and occupational therapy referrals might be helpful. Assistive devices such as a walker or wheelchair can increase safety and make it possible for people to return home after hospitalization. An assistive device can be overwhelming at first because it is a reminder that the disease is progressing. Some persons with HD will try to resist using an assistive device. However, using an assistive device can allow your person with HD to remain independent and at home for longer because it is safer and will reduce injuries.

Sudden changes in behavior could indicate your person with HD has had a head injury related to a fall.

Dr. Peg Nopoulos, a psychiatrist at the HDSA Center of Excellence at the University of Iowa, said it is “not at all uncommon for someone [with HD] to have fallen and not [remember] that it happened.”

Other signs that your person with HD might have fallen at home include furniture that is out of place, or a head lesion that can't be explained. If you suspect your person with HD has fallen, it is important to have a brain scan done to ensure there is no internal bleeding.



Often family members consult with HCP when their loved one becomes increasingly aggressive. Your safety and the safety of others in the household are important.

According to Dr. Nopoulos, “You don’t want to wait until somebody gets hurt. [If] behavior is getting out of hand and there’s concern that there’s a risk to anybody, that’s when you consider hospitalization.”

## **When Your Person with HD is Resistive to Treatment**

Persons with later stage HD sometimes resist medical treatment. It can be difficult to get them to leave the home for any reason. It might be helpful to plan ahead and explain a reason for going to the hospital that is most acceptable to them. For example, you might say that you are going to have medications updated or talk about a topic that feels less threatening to your person with HD. If you are concerned about your safety at home, and your person with HD needs to be admitted for psychiatric treatment immediately, call 911. Explain to the dispatcher and to the emergency responders when they arrive that your person with HD has HD and is aggressive. Explain that HD involves motor impairment and cognitive and behavior changes so they are prepared to manage them. Many states have a Crisis Intervention Team (CIT) that is equipped to handle mental health emergencies.

If there is not an immediate emergency, but your person with HD’s behavior is getting worse and they are resistive to treatment, it might be necessary to arrange an inpatient psychiatric hospital stay.

Psychiatric commitment hospital stays are common. Families sometimes feel that this action is extreme, but according to Dr. Nopoulos “it is actually the best way to take care of the person. For example, if your loved one is psychotic and not taking their prescribed medications, a psychiatric commitment may be the only way for the person to get the proper treatment.” She continues by saying that even though it seems dramatic to force a mental health stay, mental health commitments are extremely common and sometimes “it’s the only way that we can treat the person. In many ways it’s the most important thing to be done.”

In every state a law enforcement officer is empowered to detain an individual and remove them for a mental health evaluation. The process differs in every state. You will certainly be asked if your person with HD is at risk of harming themselves or others. If you are able to get your person with HD safely to the hospital, the physician at the hospital can initiate the commitment process. This might be more comfortable for family members to not be the ones seeking a commitment if it can be done safely. It would be helpful to familiarize yourself with your state’s commitment laws so that you understand the process of seeking assistance for your person with HD.

[http://treatmentadvocacycenter.org/storage/documents/Emergency\\_Hospitalization\\_for\\_Evaluation.pdf](http://treatmentadvocacycenter.org/storage/documents/Emergency_Hospitalization_for_Evaluation.pdf)

## **Plan Ahead**

It’s important to have a plan in place ahead of time regarding what types of interventions are acceptable to you and your person with HD when you go to the hospital. Advance directives are legal documents that specify a person’s preferences for medical treatment at the end of life and designate a person to make healthcare decisions for them if they are no longer able to decide for themselves. States vary in how these forms are prepared. Go to the National Hospice and Palliative Care website ([caringinfo.org](http://caringinfo.org)) for information about how to prepare documents in your state. In some states the document about patient preferences—also called a living will—and the document specifying a healthcare or medical power of attorney are separate documents; in other states they can be combined into one advance directive form. For assistance in creating advance directives for a person with HD, see HDSA’s Family Guide Series booklet, “Advance Directives for Huntington Disease” (you can order copies or download this resource or at [hdsa.org/shop/publications](http://hdsa.org/shop/publications)).



Advance directives are best created while the person with HD is still able to make decisions about their preferences for medical treatment. Having advance directives can help to avoid conflicts between family members regarding a person's end of life wishes. It can also ease feelings of doubt and guilt later for the person making healthcare decisions if they are following the wishes of their loved one.

Some important health issues to include in advance directives for a person with HD:

- Who will be the designated healthcare or medical power of attorney,
- Whether or not to place a feeding tube when choking becomes more frequent and dangerous,
- Whether to place a ventilator if breathing becomes difficult,
- How aggressively to treat non-HD conditions such as cancer or heart failure,
- Whether to try to avoid hospitalization at end of life.

It can feel uncomfortable having conversations about end of life; however, everyone, regardless of their health status, should have advance directives. For information on how to talk about this issue with your person with HDs, visit the Start the Conversation website ([theconversationproject.org](http://theconversationproject.org)). This website has a starter kit to help you begin talking about this topic with loved ones and will guide you through the process. Be sure to include children who are adults or reaching adulthood in these conversations. They might also be asked to make healthcare decisions for your person with HD in your absence.

Conversations about advance directives should occur every year and whenever major changes in health status occur. Some people suggest this conversation take place around the person's birthday or at annual HD-related clinic visits. These conversations should include your person with HD's HCP and you should provide your HCP with updated documents if they change. At some point your person with HD will no longer be able to participate in these conversations. A neurologist or neuropsychologist can perform cognitive tests to determine when your person with HD is no longer able to make decisions independently. At that time, the person designated as the healthcare power of attorney in the advance directives will be in charge of making healthcare decisions for your person with HD.

Even if you have a plan in place, it can be difficult to make these decisions when your person with HD is suffering. You and your person with HD always have the option to change your mind about what you want. However, a predetermined plan is helpful when decisions have to be made quickly and can help family members feel more confident that they are following the wishes of the person with HD.

## **What to Expect If You Go to the Hospital**

If you do decide to go to the hospital, be prepared for your person with HD to receive medical intervention. Some possible medical interventions might include placing a breathing tube or a feeding tube, or administering IV fluids or medications or giving antibiotics. If your person with HD is agitated or aggressive treatment might include psychiatric medications to control behavior, or the use of physical restraints. The medical team might also order diagnostic tests such as a head CT or MRI if your person with HD has had a fall. Try to anticipate what interventions might occur and be prepared to advocate on your person with HD's behalf. Have a copy of the advance directives to give the medical team. Although you might have provided a copy of the advance directives to the hospital already, it's best to bring another copy of each with you to ensure they are readily available.

If you call an ambulance to transport your person with HD, be prepared to explain to them something about HD when they arrive. Since HD is a rare disease, many emergency care workers have not had experience taking care of someone with HD. You can have a sign ready to place on the front door with brief information about HD if the emergency is not immediately life-threatening. The sign can include a picture of a STOP sign symbol that states "Read before entering: This patient has Huntington disease. This is a neurological disorder that affects movement, thinking, and behavior."



Keep in mind that many HCP have little or no experience caring for persons with HD. Be prepared to be actively involved in your person with HD's hospital care. Dr. Nopoulos reminds family members how important it is to be proactive with the medical team who may not understand that the less obvious cognitive problems may be of the most concern. She says "don't wait for them to call you....Call the nurse and say you want to talk to the doctor. This is your right even if you don't have power of attorney or guardianship. You're the one who knows the person best, and the care team needs that information from you, in order to take the best care of the person."

One important piece of information that is important to communicate to the care team is your person with HD's level of functioning. Inform nursing staff about your person with HD's level of independence in walking. If they use an assistive device for walking, make sure to bring it to the hospital and label it with their name. If your person with HD is verbal and still able to walk independently, it might not be obvious that their judgment is impaired and they require 24-hour supervision due to impaired judgment. Many HCP might be most familiar with HD as a movement disorder. They might not have a good understanding of the cognitive and behavioral symptoms. HDSA has an excellent resource you can provide the medical team or recommend to them, *Understanding Behavior in Huntington's Disease: A Guide for Professionals* (available on the HDSA website).

Another important thing to tell the medical team is how to best communicate with your person with HD. They might not know that the cognitive changes in HD affect the ability to communicate. Some persons with late-stage HD have difficulty talking but can understand everything that is said to them. Make sure staff understand this so they are aware of what they say in your person with HD's presence and ask staff to speak directly to them. Ask them to use your person with HD's name when addressing them, sitting at eye level when possible, and to use short sentences and ask "yes" or "no" questions. If your person with HD uses an assistive device to communicate, such as a message board, provide that for nursing staff or request a speech therapy consult to provide one.

Provide nursing staff with information about your person with HD's routines. As you know, frustration that can lead to behavior issues can be avoided if your person with HD is able to anticipate what is going to happen next. This will be a challenge in the unfamiliar environment of the hospital. Encourage nursing staff to talk to your person with HD before performing any cares or assessments. Inform them that this can help to avoid any potential behavior issues that can occur due to frustration, confusion, or feeling a lack of control.

Making things as much like home as possible might help your person with HD feel more comfortable and avoid feeling frustrated. Small and familiar comforts can make a big difference. Bring one or two familiar personal items from home such as framed pictures or a favorite pillow or blanket. Make sure everything from home is clearly labeled with your person with HD's name and that staff have recorded these personal items in case your person with HD is transferred to another unit or facility. If you know your person with HD is going to be transferred, try to be there to move personal items yourself. Ask staff if there is any food or personal items in a place other than your person with HD's room that should be transferred with them.

Provide the medical team with a list of your person with HD's food preferences and food texture needs (soft food, pureed food, etc.). Let them know your person with HD's typical eating schedule and emphasize that people with HD need more calories than most persons and frequent small meals. If allowed, bring some favorite food items from home and label them. If not, ask to see a copy of the food services menu and choose food items you think your person with HD would eat. Encourage staff to order extra food on meal trays to label and put away for snacks. If your person with HD needs to eat in the middle of the night, make sure to let the staff know this.

Bring some favorite clothes and pajamas from home and let the nursing staff know what they prefer to wear at bed

time and during the day. Inform them that many persons with mid and late-stage HD prefer soft, light, loose-fitting clothing, even in colder months. Let the nursing staff know what time your person with HD usually goes to bed at night and gets up in the morning and whether it is normal for them to get up at night. Inform staff if there are any particular routines your person with HD has to get ready for bed (a favorite TV show, showering before bed, etc.) Indicate your person with HD's grooming preferences. Do they prefer a shower or bath? How often do they bathe and what time of day? Do they use a comb or a brush and prefer a particular type of soap, shampoo, toothbrush, toothpaste, etc.? Bring grooming items from home if possible and label them clearly.

If your person with HD has late stage HD and the advance directives specify they do not want to go to the hospital, this is a good time to involve a local hospice organization. You can find one on the National Hospice and Palliative Care website ([caringinfo.org](http://caringinfo.org)). A hospice organization can advocate to keep your person with HD at home and help ensure he or she remains comfortable.

## **When It's Time to Go Home**

Sometimes, persons with HD are admitted to the hospital following accidents or due to short-term illness, or a need for medication changes to control behavioral or mood issues, and the expectation is that they will be discharged back home. Make sure you communicate honestly with the medical team regarding what is happening at home. Request to speak with the social worker assigned to the unit. One of the roles of the social worker is to make discharge arrangements. Remember that the social worker might not be familiar with HD and the unique needs of persons with HD and their families. Be prepared to provide the social worker with some of the resources mentioned in this chapter that explain the needs of persons with HD. Be honest about what is happening at home and whether you are still comfortable caring for your person with HD or if it is still safe to have the person at home. The social worker can also help you find respite care opportunities and determine payment needs and resources. Even if you are not yet ready for long-term care placement, you can learn more about facilities that might be appropriate for the future. Hospitalization can also be an opportunity to have your person with HD evaluated by specialists for other issues such as eating and swallowing, communication, and mobility that are described in other chapters.

## **When It's Time to Consider Long-Term Care**

Sometimes, hospitalization can be the determining factor that it's time for your person with HD to go to a long-term care facility (LTCF). Over half of people admitted to the hospital in the later stages of HD are discharged to long term care facilities. Worsening motor symptoms, and psychiatric and behavioral issues, including aggression, are some reasons why persons with HD are admitted to LTCF. The safety of caregivers is the most important consideration. If your person with HD is aggressive or violent and you do not feel safe at home, you should consider LTCF placement. Medication adjustments might reduce aggression temporarily. Often hospitalization is the time the family and the care providers start having the conversations about whether it is time for long term placement in a facility.

Many families struggle with the decision to place their loved one in a long-term care facility. It is important to have this discussion with your person with HD long before this becomes an issue. Some persons with HD say they would prefer not to become a burden to family members when they reach late stage HD and want to be placed in long-term care. When they reach the late stage they might have impaired judgment and insight into their care needs. This might lead them to be resistant to long-term care, even if they stated this preference earlier. Although families experience guilt for wanting to place loved ones in long-term care, it can be comforting to know that their loved one at one time expressed that they did not want to become a burden to their families. Seek advice and support from members of your local HDSA support group or from HDSA to help you decide what would work best for you and your family.

Often the issue is safety. Dr. Nopoulos makes a comparison between leaving a 3 or 4 year old alone at home and

a person in later stage HD who has impaired judgment. They might appear superficially to be interacting normally, but if their judgment is impaired significantly, they are not safe at home unattended. At an HDSA COE, clinicians will do a complete cognitive assessment, even if it's early in the disease to get a baseline so the person's cognitive function can be followed over time. This will be repeated every two years or sooner if there's concern. This is very important because sometimes people with cognitive impairment can appear normal in a superficial conversation, but a comprehensive cognitive assessment can reveal significant problems. Dr. Nopoulos says that the "cognitive tests can point out deficits that are severe enough to warrant dementia." "Often times, that clinical diagnosis is a combination of not only how they do on the test, but also information from family about whether [the person's HD] thinking skills are impairing their abilities at home." If you don't have access to an HDSA COE, a local neuropsychologist can perform the cognitive tests.

Another important consideration is what resources are available at home to help with your person with HD. If it's not financially possible to have someone stay with your person with HD 24 hours a day, or if constant supervision is a burden to any family member, then you should consider placement in a LTCF. The health of all family members is important and sometimes full-time caregiving can negatively impact caregiver health. HDSA and HDSA support groups can help family members feel more comfortable about care facility placement if this becomes necessary. They can also provide information about the most appropriate care facilities for your person with HD.

Please remember that you are not alone as you think about these hospitalization issues. Talk with an HDSA social worker to get support and answers to your questions. They are there to help you process this information and help you make the best decisions for your person with HD.

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A Physician's Guide to the Management of Huntington's Disease

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Understanding Behavior in Huntington's Disease: A Guide for Professionals.

<http://hdsa.org/wp-content/uploads/2015/03/Understanding-Behavior.pdf>

HDSA Family Guide Series "A Caregiver's Guide to Communicating with Healthcare Providers"

<http://hdsa.org/wp-content/uploads/2015/03/Caregivers-Guide-to-Communicating-with-Healthcare-Providers.pdf>

HDSA's Family Guide Series "Advance Directives for Huntington Disease"

[http://hdsa.org/wp-content/uploads/2015/03/HDSA\\_Family-Guide-Advance-Directives-for-HD.pdf](http://hdsa.org/wp-content/uploads/2015/03/HDSA_Family-Guide-Advance-Directives-for-HD.pdf)

National Hospice and Palliative Care website

[www.caringinfo.org](http://www.caringinfo.org)

Treatment Advocacy Center

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