

Physical and Occupational Therapy for Huntington's Disease

A Guide for Families
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I. Introduction

When I met my first patient with Huntington's Disease (HD) nine years ago, I knew little about the disorder. The lack of available information about physical therapy and HD meant hard work for both of us as together we evaluated his deficits and developed strategies to lessen their effects. Now, many patients later, I remain in awe of the determination and perseverance that people with HD possess in their battle against the disease. Therapeutic treatment can help them in this daily struggle.

Most physical therapists (PT's) and occupational therapists (OT's) will go through their careers without ever treating a person with Huntington's Disease. For those who are given the opportunity, however, this booklet will serve as a guide to help them develop an appropriate treatment plan. Many of the suggested strategies and interventions can be performed by family members at home - without the assistance of a PT or OT.

Therapeutic services can be delivered in a number of ways. In the earlier stages, consultation with a PT and/or OT is often adequate to identify postural changes and early balance symptoms. A home program can be established with follow-up visits to monitor progress. Later, weekly sessions may be needed to assist with walking difficulties and to teach the person how to use adaptive equipment.

As moving about in the community becomes more difficult, home visits through a home care agency may be beneficial. During these visits the therapist can help to establish a safe home environment, suggest adaptive equipment, teach compensatory strategies, and provide caregiver education. In the later stages, either at home or in a nursing facility, therapists can assist with proper seating equipment, daily living skills and preventive management.

Understanding Huntington's Disease?

HD is a progressive neurological disorder affecting cognition (thinking, judgment, memory), movement and emotional control. The symptoms of HD are caused by the loss of cells in a part of the brain called the *basal ganglia*. Symptoms appear gradually, usually in mid-life, and last 15 to 20 years after onset. Although there are commonalities, there is no set pattern of symptoms and their severity can vary greatly from person to person. For some people, involuntary movement may be pronounced from the beginning, while others may have little or none. Some may experience severe emotional difficulties, while others do not. Though the pattern and severity of the disease vary, the course of HD can be divided into three stages - early, middle, late - which will serve as the basis for discussion of treatment strategies.

The Cognitive Disorder

The cognitive disorder in HD is characterized by impaired memory and executive functions, and slowed thinking. Memory is affected in the retrieval stage, that is, the person with HD has difficulty retrieving stored information. However, storing new information can also be affected.

Executive functions include organizing, prioritizing and regulating information. Impairments in this area cause difficulties in judgment, problem solving ability, logical thinking, and handling more than one task at a time. Thought processes may also be slowed so that more time is required to answer questions and to complete routine tasks.

II. Physical Therapy In Early-Stage HD

Before discussing specific problem areas, consideration should be given to overall physical condition, including cardiovascular health. With the onset of the disease come physical changes which are not readily apparent. There may be weakness of the stabilizing musculature of the upper back and trunk, affecting posture and the ability to take deep breaths. These changes often lead to a sedentary lifestyle which, in turn, results in an overall decline in health.

Often, changes in cognition and/or emotions cause these physical changes to be overlooked. A general fitness plan at this time can be significantly helpful. This type of program could be prescribed by a physical therapist and then carried out at home or at a local health club.

A typical program may consist of the following:

- Strengthening exercises for muscles involved in postural control.
- General strengthening exercises for major muscle groups of the body.
- Advanced balance activities.
- Aerobic activity to enhance cardiovascular function.

Beginning a fitness program early in the disease process not only serves to maintain physical function, but also helps to reduce some of the stress associated with catastrophic illness. A routine fitness program can also be very helpful in treating depression, which is common in HD. While a program of this type can stabilize a person's symptoms, however, it does not alter the course of the disease.

Common Motor Deficits Leading To Falls

Muscle Weakness

Muscle weakness may be widespread, depending on the person's activity level. Common areas of weakness are:

- The extensor muscles of the neck
- The postural muscles of the trunk
- The intrinsic muscles of the hands and feet

These weaknesses may lead to a forward head, rounded shoulders, or protruding abdomen. Picking up small objects with the fingertips may become difficult. Similarly, weakened foot muscles prevent the toes from grabbing the floor when balance is offset, causing loss of balance. Other consequences include shortness of breath upon exertion or difficulty taking a breath deep enough to clear secretions.

Balance

- Balance problems often begin with inability to balance on one foot. Balance is further compromised as reaction time slows. Loss of balance occurs most frequently when:
 - Turning quickly, especially when carrying a load.
 - Suddenly changing direction.
 - Wearing unsupportive footwear, such as high heels, sandals, slip-ons or worn footwear.

The combination of changes in the postural musculature that support the trunk and changes in balance leads to falls and difficulty walking. If falls are occurring infrequently, balance and strength training are most appropriate. Adding wider bases to footwear may also enhance stability.

Subtle changes in *sensation* (the ability to notice changes in the environment) and *perception* (the ability to interpret information from the senses) may also be occurring. A physical or occupational therapist can assess changes in these areas and make suggestions to prevent accidental injury. Some common problems are:

- Difficulty determining hot and cold. This can lead to burns or overexposure.
- Infection of minor cuts and scratches. Being unaware of the injury, the person does not cleanse the wound properly.
- Decreased spatial awareness. Misjudging the distance of an object, or the height of a stair or curb, can lead to falls.

III. Physical Therapy In Middle-Stage HD

As Huntington's Disease progresses the difficulties encountered in the earlier stages become more pronounced. Involuntary movements, most often chorea, may worsen and medication to suppress the movements may be helpful. It should be noted, however, that the side effects of the medication may cause increased swallowing/speech difficulties or problems with walking, and should be monitored closely. Many of the problems that chorea poses are social ones and people with HD can often manage their chorea fairly well without the use of medications.

Common Motor Problems and Strategies

Strategies and equipment can reduce the risk of injury while helping the person with HD to maintain as much independence as possible

Common Motor Problems and Strategies

Changes in muscle tone are common at this stage of the disease. This may cause the individual to feel stiff and to move awkwardly.

Common Motor Problems and Strategies

Relaxation techniques can reduce this problem.

Problem

Reduced proximal stability, or the weakness that affects the postural muscles, now prevents these muscle groups from stabilizing the back. This causes difficulty in maintaining proper sitting and standing positions.

Strategies

- Exercises and activities which encourage proximal stability can be prescribed by a physical therapist. .

- A PT can also help adapt furniture to promote improved posture.

Problem

Impaired balance causes many problems. Even sitting down in a chair requires a change in balance from one foot to the other, and for those with impaired balance, this can lead to a fall. Spatial awareness deficits can compound balance problems, making everyday tasks dangerous.

Strategies and Equipment

- *Sitting down.* Teach the person with HD to touch the furniture prior to sitting down. In a three-part process called TOUCH-TURN-SIT, the person approaches the chair and touches it (to stabilize him/herself), then turns and finally sits. This simple strategy solves the balance problem as well as the perceptual problem of being unable to judge distance.
- *Standing up.* The person should first place his/her hands on knees, bringing the weight forward and ensuring that he/she is bending from the waist. The feet should then be placed directly under the knees. The final step is to "push up" to a standing position.

NOTE: All strategies may be taught and rehearsed in the earlier stages of HD.

- *Walkers.* Specialized equipment such as a walker may be helpful. A rollator walker with swivel castors is the walker of choice, as long as the person has adequate motor control in the arms. A rollator walker with adjustable hand positions is preferable for people who have less motor control and need to have their center of gravity brought forward to reduce backward falls.
- *Safety equipment,* such as a helmet or elbow and knee protectors, can provide the necessary protection when learning new skills or trying to maintain independence. Not surprisingly, it is often difficult to convince an individual to wear a helmet or other protective gear. A soft head protector is sometimes more acceptable and can offer good protection from injury caused by falls.
- *Set up a schedule* for wearing protective headgear for very short periods each day. In this way the individual can dictate what time of day he/she will wear it and thus have some control over the decision. This will help foster acceptance and encourage regular use.

Problem

Fatigue contributes greatly to falls. Most people with HD are unable to identify fatigue. They have a tendency to continue their daily routine oblivious to changes in their gait pattern. Careful observation usually identifies changes that are caused by fatigue.

Strategies

- Some people cope with these changes by building a rest period into their daily routine.
- Others may choose to use a wheelchair for part of the day or for extended distances. They may start their day out by walking, then switch to a wheelchair when their gait pattern starts to deteriorate.

Wheelchairs and Seating

Choosing the right kind of wheelchair and other seating equipment is important.

- Many people with HD have an easier time mobilizing a wheelchair with their feet. In this case, a hemi-height or drop seat wheelchair is most appropriate so that the person's feet plant firmly on the floor.
- A small wedge cushion or lumbar support may be helpful in providing the proper support for extended sitting.
- Padding of the armrests may be necessary if chorea is pronounced. Commercially made armrest bolsters work well and provide lateral support.

The following suggestions, developed by Lori Quinn, EdD, PT, may be helpful in determining optimal seating for a person with HD:

- Keep restraint use to a minimum (see below) Allow enough room for the person to move around freely and without injury.
- Protect from hard surfaces and sharp edges with proper padding.
- Maximize ease of transfers and provide for independent mobility if appropriate.
- Make sure there is solid, sturdy foot support
Seating should be at the right height for use at the table.

Strategies To Improve Cognitive Functioning

As the motor disorder progresses, the cognitive disorder may interfere with the learning of new tasks. To compensate, some simple strategies can be employed by family members and professional caregivers alike.

To enhance the learning process:

- Break the skill down into simple steps.
- Practice through repetition.
- Allow ample time for learning.
- Provide 'cues' or "hints" either verbally or in writing, but keep feedback to a minimum and keep it goal-directed.

To enhance attention and concentration-

- * Provide a quiet environment with few distractions.
- * Make expectations clear and direct.
- * Underline or repeat key points in directions.
- * Keep materials to a minimum.

IV. Physical Therapy In Late-Stage HD

In the advanced stages of the disease, motor control is greatly diminished. Most individuals become more rigid and have little ability to change positions voluntarily. Chorea may also worsen. In addition, difficulty modulating the force of movements creates a whole new set of care and safety problems.

The same problems exist whether the person is cared for at home or in a nursing home, but many nursing homes are ill-prepared for the spectrum of problems posed by Huntington's Disease. With this in mind, the following suggestions are geared primarily to the nursing home setting, but can also be applied in the home.

Avoiding Restraints

Although restraint use has been highly regulated in nursing homes since the late 1980's, its use for people with HD continues to be commonplace. Unfortunately, restraints tend to create more problems than they solve. A common use of a restraint is a seat belt or waist restraint to prevent falling out of a chair. What is overlooked is the reason *why* the person is falling out of the

chair. Often uncontrolled movements can be minimized by a change in position or improved seating.

In many cases, specialized seating needs are not taken into account. For example, a chair with a high back, increased seat depth, foot support, armrests and padding (if chorea is present) may be necessary. Chairs that have been designed with these features in mind are commercially available (see product information in the Appendix).

The person who has impaired balance, impaired self-awareness, spatial perception deficits and impulsivity poses the most problems for the caregiver. This is the individual who appears never to sit still, is constantly attempting to stand up and falls almost immediately each time. In order to provide a safe environment for him/her, the family or staff must establish a consistent routine based on the person's wishes and needs. This may involve:

- Several opportunities for the person to walk with assistance throughout the day.
- Frequent position changes. Frequent meals.
- Chair and bed alarms to alert the caregiver to any unsafe behavior.

The Sleeping Environment and Bedding

Sleeping in a regular bed can be difficult for many people with HD. Decreased spatial awareness may reduce ability to sense the edge of the bed during sleep, causing the person to fall out of bed. Inability to modulate force of movement may also cause the person to 'vault' out of bed when he/she simply wanted to turn over or sit up.

In the home setting, placing a mattress on the floor and clearing the surrounding area of any furniture often is sufficient to create a safe sleeping environment. In the nursing home, however, this is not always possible. Side rails fixed to the bed are frequently tried as a solution, but more often than not they become obstacles to climb over or bang up against.

People who are still able to rise to sitting and standing tend to do best with a very low bed three to five inches off the floor. A thin, high density mat can be placed on the floor at the side of the bed to cushion a fall should it occur. In some cases, one side rail can be used with at least two inches of foam padding to provide a sensory boundary, leaving the other side open for entering and exiting the bed.

In extreme cases of inability to modulate force of movement, a more confining arrangement is required to prevent serious injury. Unfortunately, commercially available options are few. A bed enclosure which fits over a regular hospital bed can provide a see-through method, but many people report feeling too confined (see product information in Appendix).

Another alternative is to build a modification of a Craig bed. A platform the size of a full mattress is built eight inches off the floor with four-foot high walls which are padded with foam and covered with smooth vinyl. One side of the bed opens completely for ease of transfers and care. One drawback is the appearance of isolation, but the opportunity of a good night's sleep, the ability to change position without the risk of falling, and the opportunity to sit up without crashing into a side rail, usually outweigh the drawbacks.

V. Occupational Therapy In Early-Stage HD

As intellectual changes begin to occur, an occupational therapist can help the person with HD and family members develop compensatory strategies. By employing these strategies early in the disease process, the person with HD learns that he/she can have some control over the disease and is therefore less apt to feel helpless.

Memory Strategies

- Offer hints, such as word associations, to aid in the retrieval of information.
- Use lists, calendars and notes.
- Establish a consistent daily routine.
- Label items around the house.
- Avoid open-ended questions. Instead, offer a list of choices and ask questions requiring a "yes/no" answer.

Difficulty in sequencing tasks is another common problem experienced in the early stages. Family members report that tasks that used to be completed in a few hours now take all day or are never completed at all. The strategies listed below can help to remedy this problem.

Planning Strategies

- Write down all tasks in a logical order.
- Review the steps to ensure that they are clearly understood.
- Break complex tasks down into parts.
- Encourage completion of each step prior to moving on to the next step in the task.

Concentration Strategies

- Use short sentences when giving directions.
- Ask the person to repeat important points back to you.
- Reduce outside stimuli whenever possible.
- Underline key points in directions or recipes

Safety in the Home

A home consultation by an occupational therapist can identify unsafe practices and provide suggestions to improve safety. Analysis of meal preparation skills, personal hygiene, and other activities of daily living (ADUS) can lead to strategies which foster independence. Here are some examples:

In the Kitchen

- Use unbreakable dinnerware.
- Avoid storage of commonly used items in high cabinets.
- Use a kitchen timer as a reminder to turn off appliances.
- Lower the hot water temperature to prevent scalding.
- Use covered mugs for hot liquids.
- Use oven mitts rather than pot holders.

In the Bathroom

- Use a non-skid mat in the tub or shower.
- Use 'soap on a rope'.
- Use a shower bench or chair.
- Use safety bars.

In the Bathroom

- Stabilize furniture so that it cannot move.
- Use chairs with high backs and armrests.
- Clear rooms of any unnecessary furniture.
- Remove scatter rugs or thick pile carpeting.
- Keep tables and lamps away from walkways.
- Pad doorways and furniture when contact is frequent.

The combination of motor control problems and cognitive creates a number of difficulties in activities of daily living. Again, strategies and equipment can be of great use in prolonging independence.

Eating

Problem

Impaired postural control creates positioning problems at the table. Slouched sitting and inability to maintain proximal stability cause a great deal of food spillage as well as fatigue.

Strategies

- Have the person sit in sturdy chair as close to the table as possible.
- He/she should wrap legs around legs of chair to stabilize the pelvis and put elbows ON the table to stabilize the upper trunk.

Problem

Motor impersistence and muscle weakness cause difficulty holding onto utensils, or difficulty bringing hand to mouth

Strategies

- Use utensils with built-up handles.
- Utensils should be put down after each bite to rear muscles and prevent fatigue.

Problem

Incoordination of movement makes simple tasks such as cutting food seem very difficult.

Strategies

- Use non-skid placemat to prevent dishes from moving.
- Use covered cups or mugs (travel mugs are ideal) to prevent spills.

Hygiene

Problem

Difficulty with motor planning and sequencing tasks can make even the most routine activities seem insurmountable. Though some people with HD lose interest in personal hygiene as a result of depression, this apparent lack of interest often is a result of diminished planning and sequencing ability.

Strategies

- Divide morning and evening routines into simple steps.
- Write out the steps for completing all hygiene and posts in a visible place.

Problem

Maintaining balance while performing everyday tasks, such as shaving or brushing teeth, can become difficult and even hazardous.

Strategies

- Use a shower bench or chair to prevent fatigue and assist with balance.

Problem

Combining a fine motor task, such as holding the soap, with a gross motor task, such as washing the torso, also presents difficulties.

Strategies

- Use shower mitt (the soap can be put right inside the mitt so it won't fall out) or "soap on a rope".
- Use an electric razor or chemical hair remover.
- Build up handle on toothbrush or hairbrush by wrapping and taping a washcloth around it.

Problem

Fatigue often plays a role in preventing the completion of tasks.

Strategy

- Build rest periods into the routine.

Dressing

Problem

Decreased decision-making ability may cause people with HD to avoid changing their clothes because they have difficulty deciding what to wear..

Strategies

- As with other complex tasks, reduce the routine to simple steps.
- Label and pair favorite items so that decision-making is kept to a minimum.

Problem

As the small muscles of the hands weaken, ability to grasp objects is diminished.

Strategies

- Avoid clothing with multiple fasteners.
- Put a ring on zippers to aid in opening and closing.
- Encourage dressing while sitting in a sturdy chair to reduce falls and fatigue.

Problem

Lack of initiation - the inability to start a task - may necessitate a 'jump start'.

Strategy

- Suggest an activity or task and offer to help. Often this will provide the necessary impetus for the person to complete the task independently.

VII. Occupational Therapy In Late-Stage HD

In the later stages of the disease, preventing injury to the body becomes the utmost concern. Although people with HD appear to be moving all the time, they are often not able to change position voluntarily, and are therefore susceptible to skin breakdown from constant pressure or skin tearing from constant shearing movement.

The occupational therapist can assist the caregivers by suggesting a routine of position changes. The OT can also assist in designing a protective environment for the person with a lot of choreic movement. Padding of hard furniture, wheelchair parts and sharp corners helps to prevent injury from falls or choreic movements. In cases of very severe chorea, it may be necessary to pad a part of the body if constant contact is being made.

Because the person with advanced HD is no longer able to control movement, certain muscle groups are no longer used. This disuse, combined with changes in muscle tone, can lead to permanent disability called *Contracture*.

Contractures

A contracture is the permanent shortening of a muscle. Contracture management in HD can be difficult because of fluctuations in muscle tone and the presence of chorea. Frequent position changes and range of motion exercises are important weapons in the battle against contractures.

In some cases splinting can be helpful. New air-assist-type splints, which use air bladders to provide support and have enough "give" in them to avoid skin breakdown, have proven very helpful in the management of elbow and knee contractures. Foam core hand splints have also proven useful for maintaining functional positioning of the hand. These are very lightweight and have a washable cover for easy care (see product information in the Appendix). Many people are able to wear splints during the night. This gives them eight to ten hours of appropriate positioning and slows down the contracture process.

VIII. Conclusion

Today, more than ever, there is hope for people with Huntington's Disease. Rapid progress is being made toward potential treatments for HD. Likewise, quality of care for people with HD has improved greatly in recent years. By seeking early intervention from health professionals such as physical, occupational and speech- language therapists, the person with HD can have control over the disease and over his/her life. By learning strategies to help them cope with the many changes that are brought on by Huntington's Disease, people with HD can live meaningful, productive lives well into the final stages.

IX. Appendix

How to locate an Occupational or Physical Therapist

1. Physician referral.
2. Contact the Huntington's Disease Society of America, 158 West 29th Street, 7th Floor, New York, NY 10001-5300, 800-345-HDSA, 212-242-1968 for a referral.
3. Call any major hospital. They should have a rehabilitation department which includes physical and occupational therapy.

Product Information

Walking Devices -Rolling Walkers, Standard Walkers, Canes

Sammons Preston, P.O. Box 5071
Bolingbrook, IL. 60440-5071, 800-848-WALK

Strider Walkers

Guardian Products, 4175 Guardian Street
Simi Valley, CA 93063, 800-423-8034

Safety Devices - Helmets, Elbow/Knee Pads

Best Priced Products, P.O. Box 1174
White Plains, NY 10602, 800-824-2939

Sammons Preston, P.O. Box 5071 Bolingbrook, IL 60440-5071, 800-323-5547
J.1 Posey Co., 5635 Peck Road
Arcadia, CA 91006-0020, 800-44-POSEY

Chairs

Broda Seating, 385 Phillip St, Waterloo, Ontario NGL SR8, Canada, 800-668-0637
Gunnel Inc., 8440 State Street Millington, MI 48746, 800-551-0055
Hill Rom, 1069 State Bank 46 East Batesville, TN 47006, 800-445-3730

Huntington's Tilt in Space/Recliner

Gunnel Inc., 8440 State Street
Millington, MI 48746, 800-551-0055
Hill Rom, 1069 State Bank 46 East
Batesville, TN 47006, 800-445-3730

Positioning Devices - Cushions, Supports, Padding

Alimed, 297 High Street
Dedham, MA 02026-9135, 800-225-2610

Lumbar Supports

J.T. Posey Co., 5635 Peck Road
Arcadia, CA 91006-0020, 800-44-POSEY

Eating Utensils, Non-Skid Matting, Shower Benches and Chairs, Shower Mitts

Alimed, 297 High Street
Dedham, MA 02026-9135, 800-225-2610

care spoons	covered mugs
sure grip	Long-handled brush
plate guards	non-skid dinnerware
shower mitt	shower bench/chair

North Coast Medical
2509 E. Thousand Oaks Blvd., Suite 430
Thousand Oaks, CA 91362, 800-634-4351

care spoons	covered mugs
sure grip	Long-handled brush
plate guards	non-skid dinnerware
shower mitt	shower bench/chair

Posey Lateral Stabilizing

Smith & Nephew Rolyan
N104 W13400 Dornges Bay Road
Germantown, WI 53022, 800-521-6664

Splinting Devices - Splints For Hands, Knees, Elbows

Medassist - OP, Inc., P.O. Box 758
Palm Harbor, FL 34682, 800-521-6664

Bedding - Padding, Low Beds, Bed Enclosures

Vail Products, 235 First Street
Toledo, OH 43605, 800-235-VAIL
NOA Medical Industries, 1601 Woodson
St. Louis, MO 33114, 800-368-2337

For Further Information

Additional publications about physical and occupational therapy, nutrition, speech and swallowing, and related topics may be obtained from the Huntington's Disease Society of America,
158 West 29th Street, 7th Floor, New York, N-Y 10001-5300, 800-345-HDSA, 212-242-1968.

Visit our site on the World Wide Web: www.hdsa.org