

People with HD present with many different physical, social and psychological problems that may make it difficult for them to carry out their activities of daily living. However it is vital that their independence and mobility is maintained for as long as possible and this may require carers, relatives and or therapists to assist them to move. The following is a list of tips for handlers and the person with HD to facilitate independence and help them move more efficiently.



Table 1. Tips to assist moving and handling a person with HD

Advice	
Take Your Time	<p>People with HD are often slow to start and complete a movement, for example, when standing up from sitting. The time it takes for the brain to instruct the muscles to perform the action is slowed in HD.</p> <p>Therefore they need time and patience to allow them to do as much as possible themselves.</p>
Technique	<p>Using the technique that is most effective for the individual during any task will allow the person with HD to do as much as possible themselves, thereby increasing their independence and reducing the risk for carers.</p>
Verbal Cueing	<p>For some people with HD, counting “1,2,3, Go”, or the use of “Ready, Steady, Go” may assist them to start off the movement. Instruction during or just prior to the task may also be helpful to remind the person of the most effective technique. This will require practice overtime.</p>
Manual Cueing	<p>For some people with HD, physical assistance maybe required from the handler. This may involve a simple indication of how to achieve the most effective technique or to simply provide a point of stability for balance during the task. Of course the handler should not put themselves at risk while assisting the person with HD to move.</p> <p>Some people who have choreic movements may actually be hindered by hands-on assistance.</p>

Table 1 continued. Tips to assist moving and handling a person with HD

Advice	
Simple Commands	<p>Use simple short commands to request a task. This is especially true for people with any cognitive/ understanding difficulties.</p> <p>For the person with HD, too much complicated explanation or too much repetition of a simple request will interfere with the brain's ability to process the information and will ultimately slow the ability to perform the task further.</p>
Practice	<p>The person with HD will benefit from repetition of simple tasks during the day/ week. This is especially true if they have not been doing the task themselves for some time.</p>
Consistency	<p>It is vital that all handlers (therapists, carers, relatives) involved with the person with HD are assisting in the same way. This will increase the amount of practice the person gets and therefore improve their proficiency of the task.</p>
Motivation	<p>Apathy and depression are common symptoms associated with HD and often result in the person lacking the motivation to move regularly. Encouragement to move and positive feedback during and after tasks is important.</p> <p>One way to motivate is to set simple achievable goals that are important to the person with HD. This could involve the completion of a simple task independently, within a set time (e.g., complete 3 sit to stands in 60 seconds), over a set distance (e.g., walk 20 metres in 60 seconds), or may involve a fun element (e.g., hold a tray and balance a tin of beans on the tray while standing up from sitting).</p>
Be Calm and Reassuring	<p>Anxiety and irritability are symptoms of HD. If these symptoms are increased during movement, the handler could try a distraction or simply change the task to avoid the behaviour escalating.</p>
Equipment	<p>Simple pieces of equipment can be very useful in allowing the person with HD to move independently. For example, grab rails and sliding sheets.</p>

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FOOTWEAR

Wearing appropriate footwear is important because shoes can make a dramatic impact on what an individual is able to do. They provide a foundation for posture. Upright stability is important because it affects balance and walking ability. There are certain features to look for in a shoe that can help to maintain balance and decrease the risk of falling. As shoes can wear down very easily in people with HD, it is therefore important to recommend regular checking of footwear and purchasing new shoes as needed. The following is a list of what to look for when choosing footwear to promote safety and function:

- firm, flat heel
- bendable forefoot
- velcro shoelaces for ease of application
- wide heel base for increased stability
- firm heel counter that keeps the heel supported and prevents oversupinating/overpronating
- avoid thick soles
- avoid thick toe grips (thick toe grips and soles can catch and lead to falls)
- high top sneakers/shoes recommended for ankle support
- avoid flip-flops and high heel shoes

The following footwear meet the recommended criteria:



ORTHOTICS

Although orthotics are not often prescribed for people with HD, they may be beneficial.

Arch supports and orthotics worn inside of a shoe that position the foot in subtalar neutral may improve ankle motion and lower limb stability.

If ankle dystonia is a problem, a heel wedge and/or lateral wedge may improve ankle movement in the inversion/eversion direction and an ankle foot orthosis (AFO) may improve movement in the dorsiflexion/plantarflexion direction.

A custom made shoe inlay may also be helpful for individuals with clawing of toes during walking.

Lateral wedge to
prevent excessive
supination



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Mobility and walking impairments are a major concern for people with HD. They may have choreic movement (abnormal involuntary movement caused by brief, irregular muscle contractions) which if present in the upper limbs, trunk and lower limbs may affect their gait pattern and result in decreased balance and coordination skills. They can also be affected by motor symptoms, which cause slow initiation of movements and slow, wide based gait pattern. It is vital that independence and mobility is maintained for as long as possible.

KEY RECOMMENDATIONS

FOR ALL AMBULATORY INDIVIDUALS:

- Focus on balance and safety and do not let involuntary movements from chorea influence your judgement regarding that person's ability to walk.
- Train family/carers to reinforce use of devices or any safety suggestions, such as big steps when turning or keeping hands free for balance.
- Have carers remind the person with HD to avoid dual-tasks if this is an issue for them (i.e., not to carry anything or talk when walking).
- Facilitate access to re-assessment/treatment as mobility/compliance changes.
- Encourage the use of appropriate footwear (see HD Clinical Tips: Footwear and Orthotics, for further recommendations).


FOR PEOPLE WHO FALL / AT HIGH FALL RISK:

- Create a limited, safe area to allow ambulation, especially in those with repeated falls.
- Use protective helmets/hats, elbow pads, knee pads, and hip protectors in people with HD who fall frequently.
- An individual assessment of walking devices by a physiotherapist is recommended. Although canes or walking sticks may be useful early in the disease process, once falls occur, a four-wheeled rollator walker is highly recommended.
- It will be easier for the person with HD to learn to use a walking device earlier in the disease process, thus early introduction is recommended even if the device is only used sporadically.
- Later in the disease, people with HD are often unable to use assistive devices due to cognitive issues, and may do better walking hand in hand with one or two carers.

WALKING DEVICES

Types	Considerations
 <p>Straight canes or Walking stick/s</p>	<ul style="list-style-type: none"> • Supports only small % of body weight and cannot prevent falls, but may improve balance in those with mild balance deficits. • A heavier stick may help with coordination. • Walking sticks are often preferred by people with HD as they allow natural arm movement and do not slow gait. • It is important to consider the problems associated with choreiform movements and whether sticks are appropriate.
<p>Four-wheeled Rollator Walker</p> 	<ul style="list-style-type: none"> • It is easier to push walkers/frames rather than lift canes as canes increase stance time (1). • Larger wheels move better on uneven surfaces indoors and out. • Depending on the size, walkers can be cumbersome and difficult to manoeuvre through some areas. <p><u>Modifications to consider:</u></p> <ul style="list-style-type: none"> • If it has a seat it is only safe with a back support. Otherwise modify it so that either the seat has a tray or a back. • In individuals who fall backwards, use of a heavier framed walker may be beneficial in adding additional stability. <p><u>4-wheeled frames:</u></p> <ul style="list-style-type: none"> • The 4-wheeled rollator has been shown to be the safest walking device for people with HD (1). • The 4-wheeled walker produced safer and less variable gait when compared to other devices (1).

WALKING DEVICES continued

Types	Considerations
<p>Four-wheeled Forearm Support Frames</p> 	<p><u>Function:</u></p> <p>To give more support through the forearms, particularly if hand function is impaired.</p> <ul style="list-style-type: none"> • May be cumbersome. • May provide opportunity to maintain or extend mobility and independence. • Supervision or assistance may be required. • Hard surfaces under supports may require padding to prevent self-injury from involuntary movements. <p><u>Types:</u></p> <ul style="list-style-type: none"> • Gutter frames • Split-level forearm support • Frame with single padded support.
<p>Frames with Two Front Wheels and Two Rear Feet</p> 	<ul style="list-style-type: none"> • Difficult to manoeuvre during turns; requires the user to lift while turning, which leads to more stumbles and increases fall risk. • Useful if the 4-wheeled rollator walker tends to 'run away' from the person using the walker, or when brakes cannot be used. • Best used by people who live in purpose built homes where there are no long hallways and no need to make turns. • Difficult to move over carpet pile.

KEY RECOMMENDATIONS

AMBULATION AND GAIT TRAINING

- People with HD should be encouraged to engage in regular walking throughout all stages of the disease.
- Gait training should be functional and patient-specific.
- Falls should not prevent walking (see Policy for Falls Management in Huntington's Disease).
- Footwear should enhance, not hinder walking.
- Other factors other than HD-related symptoms need to be considered when implementing gait re-training (e.g., musculoskeletal, cardiovascular systems).

WALKING AIDS

- Walking aids should enhance walking, and not just prevent falls.
- Level of support required by people with HD can be variable day to day or week to week.
- 4-wheeled walker is the walking aid of choice.

References:

1. Kloos et al 2012 *The impact of different types of assistive devices on gait measures and safety in Huntington's disease. PLoS ONE* 7(2): 1-7.

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SEATING AND POSITIONING

For Physiotherapists and Healthcare Professionals Working with People with HD

Persons with HD, especially in the middle and later stages and often spend a majority of their day sitting. Although they may be able to walk independently or with some assistance, most have at least some difficulty with achieving a comfortable sitting position. They may have difficulty sitting in a chair with inadequate back and side support and many people with HD tend to slide down in their chairs, thus maintaining weight bearing through their lumbo-sacral spine as opposed to through the ischial tuberosities, buttocks and thighs. People with choreic movements may be prone to injuries if their chair is not properly padded. Effective positioning with the appropriate supports will enable people with HD to better interact with their environment and have an improved quality of life. It is especially important to maintain an upright position especially for feeding and drinking to minimise risk of aspiration in the later stages of the disease.

Specialised seating needs should be considered; this may include increased seat back height and depth, tilt and appropriate foot support. Hard surfaces and edges of assistive devices and wheelchairs should be protected with padding where necessary. Choosing the right kind of adaptive equipment is a collaborative process. Balancing independence and safety requires special consideration for each person's individual needs. Use of certain devices and equipment, such as those described above, may provide the necessary support to maximise a person's functional abilities.

Considerations for optimal seating include:

- chair measured to ensure the correct depth, height and width for the individual
- appropriate height for use at table or with lap tray
- protect from hard surfaces and sharp edges with proper padding
- maximise ease of transfers, and provide for independent mobility if appropriate
- solid, sturdy foot support
- minimal use of restraints

Progression from independent ambulation to using a wheelchair in daily life can be very traumatic for the person with HD. This is a sign of their continued functional decline. Recommending a wheelchair as a primary means of mobility should therefore be approached cautiously. This decision should be reached with the consent of the patient, the family and the interdisciplinary team and focussing on allowing the person with HD to make their own choices to the extent that their safety and the safety of others are maintained. If hoisting for transfers, walking hoists for those able to partially weight bear are useful. When dependent for transfers, tracking hoists are usually safer as there is less risk of injury from banging against any hard parts of the hoist. Slings need careful assessments for skin protection and for the correct position to be maintained during transfers.

Table 1. Seating-related problems and possible solutions for persons with HD


Problem	Possible Solutions	
Sliding down in chair (shearing of skin can occur as a secondary problem if sat on non-slip material)	<ul style="list-style-type: none"> • The range of movement of hips, knees and ankles and also the length of the hamstrings need to be maintained to maintain/improve seating position. Use an appropriate cushion to wedge the seat, so that the seat to back angle is decreased: adjust the angle at the hip (thigh-trunk segment) in conjunction with a tilt in space chair if appropriate. • Cushion: needs to have the appropriate rating for skin-care and to provide adequate support. An ischial cut-out to anchor pelvis in place could be considered. Check that the cushion seat length is not too long. 	

Table 1 continued. Seating-related problems and possible solutions for persons with HD

Problem	Possible Solutions	
Poor postural stability	<ul style="list-style-type: none"> Chair measured for the individual. A contoured seat with/without a pommel. Lateral trunk/thoracic supports padded both sides. Tilt-in space chair, possibly with a reclining back as well. To secure the position of the pelvis use a thick, padded harness/belt, 4 point pelvic strap or groin straps. A lap tray or table to provide upper body support. 	
Bruises on arms/legs	<ul style="list-style-type: none"> Use a chair with the least amount of chrome or metal exposed as possible. Pad thickly to cover any exposed metal or hard surfaces on the chair, including the underneath of the tray; wrap the leg rests with padding. If feet tend to move off footplates, a wide calf strap may be adequate to prevent the feet catching in the front wheels, however a padded footbox provides more protection. Choose a chair which is not too confining and allows room for movements. In the later stages, conforming wheelchairs with foam carved seats contoured for the individual giving postural support/sensory input, often quietening the chorea but still permitting movement. 	 
Falls/leans to the side	<ul style="list-style-type: none"> Provide padded lateral supports at head and trunk; padded lateral supports for the hips and thighs. Recline the back. Adjust the tilt of the chair. In late stages there are definite advantages to having custom made wheelchair to provide good postural support. 	 

Table 1 continued. Seating-related problems and possible solutions for persons with HD


Problem	Possible Solutions	
Unable to tolerate upright chair Falls out of standard chairs	<ul style="list-style-type: none"> • Use a maximally adjustable chair with recline and tilt. A contoured seat with/without aommel. • Change positions frequently (e.g., 20 minutes upright, 20 minutes fully reclined). • Use pillows, padding to provide extra support/protection. • Consider the use of a bean bag chair, hammock, or padded floor bed. 	 

Table 2. Wheelchairs and seating systems for consideration

Standard Wheelchairs
<ul style="list-style-type: none"> • The standard wheelchair (self-propelling or transit) can be modified with a seat cushion to promote better sitting posture or padding for the armrests and footrests to prevent bruising. For proper support during extended seating a lumbar roll may be beneficial.
Modifications to standard wheelchairs
<ul style="list-style-type: none"> • Padding for the armrests & footrests to prevent bruising. • Spoke guards to cover the wheel spokes.
Reclining Wheelchair
<ul style="list-style-type: none"> • The back of the chair moves to allow opening/closing the angle at the hips. • Provides ability to rest from upright position, but can encourage patient sliding down in a chair.
Dynamic seating systems
<ul style="list-style-type: none"> • Tilt-in-space to provide a pivoting system for a reclined, resting position whilst maintaining posture and pelvic stability, with leg and foot plate helping to maintain ankle at right angles.
Custom made indoor/outdoor wheelchairs
<ul style="list-style-type: none"> • Moulded seat and back providing the most suitable postural support. Padding on hard surfaces including the underside of the tray. Indoor/outdoor base allowing access to activities.

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