EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

Physiotherapy Guidance Document

FOR PHYSIOTHERAPISTS

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Physiotherapy Guidance Document

Aims of the guidance notes

The aims of the *Physiotherapy Guidance Document* are:

- To provide, where possible, a scientific evidence-based document to inform the optimal, individualised physiotherapeutic management of people with HD.
- To provide a written framework for the physiotherapeutic management of people with HD, in order to enable uniformity of care internationally.
- To highlight the scientific evidence for physiotherapy practice with people with HD, evaluating the effectiveness of intervention strategies presented in the literature to allow practitioners to make an informed decision regarding patient care.
- To highlight areas within practice that currently lack the scientific evidence base to inform future research.

Professional target group

These guidance notes have been written to provide information for all physiotherapists that come into contact with people with HD. Due to the complexity of the disease, it is beneficial to have some training in this area before treating people with HD. As it is a rare condition, physiotherapists may not have had the opportunity to build up experience and expertise; this document may therefore provide a starting point for those new to working with the condition. This document will also be useful for health care practitioners and physicians who require an overview of the physiotherapist's role for people with HD to ensure referral occurs in an appropriate and timely manner.

Production of these guidance notes

These guidance notes have been created through a combination of available scientific evidence and expert consensus. To uncover the relevant evidence, a systematic approach to gathering literature was employed. This involved setting inclusion/exclusion criteria, utilising search terms, combining key-words and analysing abstracts to produce relevant articles (see Appendix 1 for literature review strategy and article summaries). The available scientific evidence has been summarised, where relevant, throughout this document. Due to the paucity of scientific evidence, recommendations were also formulated based on expert consensus from the EHDN Physiotherapy Working Group (PWG). Following completion of the initial draft document, the guidance document was disseminated to all members of the PWG and other interested healthcare professionals. Amendments were made to the document based on their feedback. In a second phase of development, sections of the guidance document were reviewed in detail by subgroups of 2-3 members of the PWG. The final document continues to be a work in progress, with plans to review and update the document as new research becomes available.

1. Overview

1.1 Pathophysiology of HD

HD is a familial neurodegenerative disease, which has an autosomal dominant inheritance (Quarrell 2009; Quarrell & Cook 2004). Offspring of an affected parent each have a 50% chance of inheriting the gene mutation, and developing the disease. The gene for HD, known as IT-15 is located on chromosome four (Rosenblatt et al. 2000). In HD, the gene is expanded due to an increased number of CAG repeats (36 repeats or more) (Quarrell 2009). Because of the excess amount of glutamine units, there is abnormal huntingtin protein production. This results in cell dysfunction and eventually cell death of medium spiny neurones of the brain. The neuronal cell loss within HD occurs primarily within the caudate nucleus and putamen of the basal ganglia, an area known to play an integral role in motor function. Degeneration is also noted within the cortex and thalamus (Quarrell 2009). These changes result in the triad of motor, cognitive and psychiatric signs seen in people with HD (Table 1).

Table 1: Common Signs and Symptoms of HD throughout the Disease Stages
(Rosenblatt et al. 2000; Kirkwood et al. 2001)

Signs and Symptoms	Pre-Manifest	Early Stage	Middle Stage	Late Stage
Motor	Mild gait changes	Mild chorea Decreased rapid alternating movements Increased muscle stretch reflexes Abnormal extraocular movements	Chorea, dystonia Rigidity and spasticity Voluntary movement abnormalities Decreased coordination Difficulty holding things Balance deficits/ Falls Dysphagia/ Dysarthria	Bradykinesia Rigidity and spasticity Severe voluntary movement abnormalities Dysarthria Dysphagia Incontinence
Cognitive	Difficulty with complex thinking tasks	Mild problems with planning, sequencing, organizing, prioritizing tasks	Intellectual decline Memory loss Perceptual problems Lack of insight or self-awareness Difficulty with dual tasking	Global dementia
Psychiatric	Depression Aggression Irritability	Sadness Depression Irritability	Apathy Perseveration Impulsivity Antisocial and suicidal behaviour Paranoia Delusions or hallucinations	Delirium

1.2 Epidemiology

A review of the epidemiology of HD has revealed an overall prevalence within Europe of between 4-9 per 100,000 (Harper 1992). The average age of onset has been reported as 40.87 years and the average age of diagnosis as 44.4 years (Morrison et al. 1995). Symptomatic onset is most likely to occur between 30-60 years of age (Morrison et al. 1995; Ramos-Arroyo et al. 2005). However, it is recognised that people may become symptomatic anywhere between 1-80 years of age (Walker 2007), with studies reporting between 7-10% of cases occurring before the third decade of life (Morrison et al. 1995; Naarding et al. 2001) (juvenile HD), and between 10-37% of cases occurring after 59 years of age (Morrison et al. 1995) (late onset). Disease prevalence appears to be slightly higher in women than in men (Ramos-Arroyo et al. 2005; Morrison et al. 1995). The average number of new cases per year has recently been estimated at 4.7 per million per year (Ramos-Arroyo, 2005). The mean duration of the disease is 17 years from diagnosis to death, although this may vary greatly from 2 to 40 years (Naarding et al. 2001).

1.3 Natural course

HD is divided into four approximate stages:
pre-manifest, early, middle and late.

Pre-manifest is defined as the time before the onset of physical symptoms of HD, although a patient has had a positive gene test confirming that they will get the disease. Once patients become symptomatic, they are typically classified into early, middle and late stages based on their Total Functional Capacity (TFC) Score (Shoulson & Fahn 1979) (see Table 2). The TFC provides a gross measure of general functioning in 5 different categories. Stage I corresponds to early stage HD, Stage II-III middle stage HD, and Stages IV-V late stage HD.

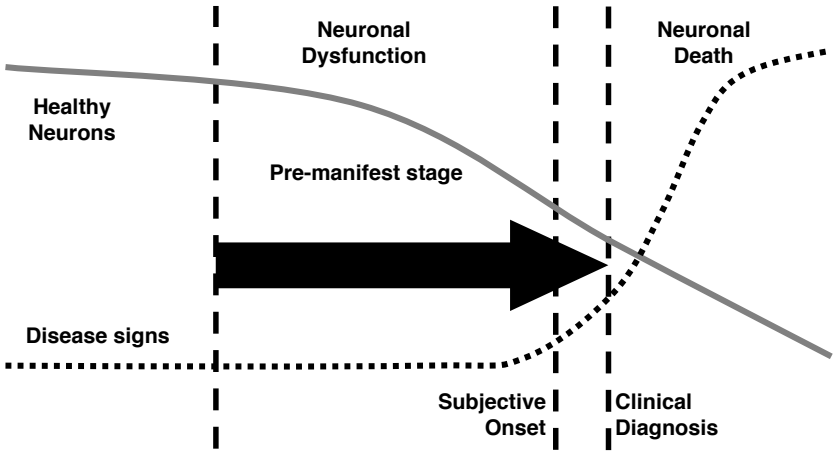
Table 2: Total Functional Capacity (TFC) Scale
(Shoulson & Fahn, 1979)

Stage of disease and associated TFC score	Category of TFC				
	Occupation	**Finances**	**Domestic Chores**	**Activities of daily living**	**Care Level**
Stage I **TFC 11-13**	Normal 3	Normal 3	Normal 2	Normal 3	Home 3
Stage II **TFC 7-10**	Reduced capacity for normal job 2	Slight assistance 2	Normal 2	Normal 3	Home 3
Stage III **TFC 3-6**	Marginal work only 1	Major assistance 1	Impaired 1	Minimal Impairment 2	Home 3
Stage IV **TFC 1-2**	Unable 0	Unable 0	Unable 0	Gross tasks only 1	Home or chronic care facility 1
Stage V **TFC 0**	Unable 0	Unable 0	Unable 0	Total care 0	Full time skilled nursing 0

1.3.1 Pre-manifest

A person who has an unfavourable genetic test for the HD mutation, but has not yet developed any clinical signs of HD (such as involuntary movements) is considered to be pre-manifest (or pre-symptomatic). In HD, the pre-manifest phase can be considered to be indicative of some neuronal dysfunction, although not indicative of cell death (Figure 1). Cognitive deterioration occurs slowly and may initially become apparent during complex thinking tasks (Rosenblatt et al. 2000). Personality and emotional changes may also be evident through uncharacteristic aggression, irritability and depression (Rosenblatt 2007a, b). Gait changes observed in pre-manifest individuals include decreased gait velocity and stride length; increased double support time; and increased variability in stride length and step time compared with controls (Rao et al. 2008).

Figure 1: The pre-manifest stage in Huntington's disease [1]



[1] Reproduced with permission from Sarah Tabrizi, Professor of Clinical Neurology at the Institute of Neurology, University College London and an Honorary Consultant Neurologist at the National Hospital for Neurology and Neurosurgery, Queen Square, London

1.3.2 Early stage (TFC Stage I)

The onset of signs of HD is insidious with no single presenting sign or symptom, and is often dependent on the sensitivity of the testing for their evaluation. Clinical diagnosis of HD, typically made by a neurologist, is currently defined by the presence of motor signs. While other symptoms, such as behavioural and psychiatric, may occur prior to the development of motor signs, motor signs are considered to be the most reliable to diagnose. These motor signs include minor involuntary movements (chorea), motor impersistence, bradykinesia (slowness of movement), deficits in voluntary saccades, and gait abnormalities (Aubeeluck & Wilson 2008; Ward et al. 1997). In addition to these motor impairments, people in the early stages of the disease may have difficulty in planning, organizing, sequencing, and prioritizing their daily activities and will complain that they just "can't get things done." Sadness, depression, and irritability are often experienced at this stage (Kirkwood et al. 2001). Within the early stages, people remain relatively functional and independent, retaining their ability to work and drive (Rosenblatt et al. 2000).

Juvenile onset HD differs from adult onset HD in that voluntary movement abnormalities, rigidity and spasticity may manifest early in the disease rather than later, and individuals have more myoclonus (i.e. sudden brief jerks involving group of muscles), and approximately 30% have seizures (Quarrell & Brewer 2009; Rosenblatt et al. 2000).

1.3.3 Middle stage (TFC Stages II and III)

During the middle stages of HD, involuntary movements such as chorea increase, dystonic postures (e.g. torticollis, opisthotonus, and arching of feet) might be present and voluntary motor tasks may become increasingly difficult (Aubeeluck & Wilson 2008). People have balance and gait deficits, including increased variability in gait parameters (e.g. stride time and length, double support time) (Rao et al. 2008; Hausdorff et al. 1998), that result in frequent falls (Busse et al 2009). Contributing factors may include bradykinesia of gait, stride variability and chorea as well as cognitive and behavioural issues (Grimbergen et al. 2008). People may also frequently drop objects that they are holding in their hands due to motor impersistence. Motor skill learning is also often impaired at this stage, resulting in difficulty learning new tasks or sequences (Heindel et al. 1988).

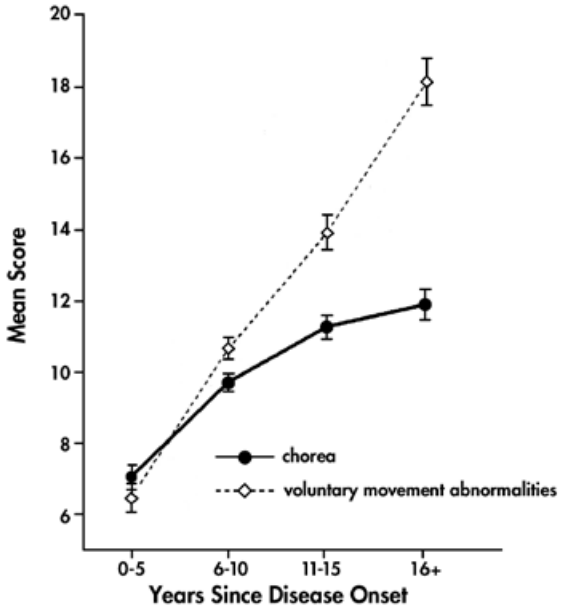
Weight loss is a common, although not universal, problem (Hamilton et al. 2004) that can begin even in the early stages of the disease. Its cause is probably multifactorial, and can be related to impaired swallowing function (Aubeeluck & Wilson 2008; Hamilton et al. 2004; Rosenblatt et al. 2000), voluntary and involuntary energy expenditure, the ability to maintain adequate energy intake, and potential abnormalities of intrinsic metabolic functioning (Hamilton et al. 2004).

Cognitive tasks become more difficult at mid-stage because of reduced ability to sequence and organise information (Rosenblatt et al. 2000). Complex types of attention are often impaired making it difficult for people with HD to do more than one task at a time (Lemiere et al. 2004). Memory loss is manifested by difficulties in learning new information and retrieving acquired information, but not in storage (Lange et al. 1995). Deficits in spatial perception (O'Donnell et al. 2008) can cause people with HD to bump into walls or tables, resulting in injuries and falls. They may not be able to recognize their own disabilities and may continue to do unsafe behaviours. Psychiatric symptoms exhibited at this stage include apathy, perseveration (i.e., difficulty with shifting to new tasks), impulsivity, antisocial and suicidal behaviours, paranoia, and delusions and hallucinations. People may no longer be able to work or drive and will need assistance when performing some activities of daily living (ADL) (Rosenblatt et al. 2000).

1.3.4 Late stage (TFC Stages IV and V)

In the advanced stages of HD, motor symptoms continue to progress, which severely limit mobility. Chorea and dystonic movement may further increase, but such involuntary movement abnormalities are often overshadowed by Parkinsonian symptoms (bradykinesia, rigidity) (see Figure 2) (Aubeeluck & Wilson 2008; Rosenblatt et al. 2000). Swallowing function can deteriorate further and people may continue to lose weight. Impaired speech results in difficulties communicating, and cognitive and psychiatric deterioration may continue, but it is thought that patients retain some comprehension (Rosenblatt et al. 2000). At this stage, cognitive deficits progress to a global dementia. Delirium, an abnormal change in a person's level of consciousness, may occur due to medications or medical problems such as dehydration and infections. At this stage most people will require assistance in all aspects of daily living, relying fully on nursing care (Rosenblatt et al. 2000). Despite the severity of the neurological disorder, the primary cause of death in HD is often pneumonia and head injury (Sorensen & Fenger 1992).

Figure 2: Changes in chorea and voluntary movement with disease progression [2]



1.4 Drug Management

Medication may be used to address behavioural or movement symptoms associated with HD. It is important for physiotherapists to know which medications a patient is on so that they can be aware of potential side effects that may affect their evaluation or intervention.

Typical medications utilised for symptomatic management of HD are summarised in Table 1 below. They can generally be grouped into four main classes: anti-choreic, anti-psychotic, anti-depressant and antiepileptic. This table is provided for information only and dosages are deliberately not provided; it is beyond the scope of physiotherapy practice to be involved with drug management. Any issues relating to drug management should be addressed by the neurologist or clinician responsible for the medical management of that patient.

[2] Progression of movement abnormalities in HD. Voluntary movement abnormalities and chorea are measured by the Quantifed Neurological Examination, mean score +SE. Higher numbers indicate great dysfunction. Scores include repeated observations of the same patient during a given time interval. The number of observations varies from 132-278 for each point. From Ross C, Margolis, R, Rosenblatt, A, Ranen, N, Becher, M, Aylward, E. Huntington disease and the related disorder, dentatorubral=pallidoluysian atrophy (drpla). Medicine. 1998; 76: 305-338.

Table 1: Main drug classes, examples [3] and potential side effects

Class of Drug (main action) (Bonelli et al. 2004; HDSA 2000)	Subclass of Drug	Example Medications	Potential side effects
Antichoreic	**Neuroleptics and atypical antipsychotics**	Tiapride (Tiapridex, Synthelabo) Fluphenazine (Prolixin) Risperidone (Risperdal) Olanzapine (Zyprexa) Pimozide (Orap) Geodon (Ziprasidone)	drowsiness, apathy, extra-pyramidal symptoms, dystonia, akathisia (restlessness), hypotension, dizziness, headache, insomnia, constipation, dry mouth, weight gain, tardive diskinesia
	Benzodiazepines	Clonazepam (Klonopin); Diazepam (Valium); Temazepam (Restoril)	sedation, ataxia, apathy, withdrawal, seizures, fatigue
	Dopamine Depleting Agent	Tetrabenazine (Nitoman)	hypotension, drowsiness, depression, gastro-intestinal disturbance, extra-pyramidal symptoms
Antipsychotic		Olanzapine (Zyprexa)	drowsiness, apathy, akathisia (restlessness), hypotension
Antipsychotic		Haloperidol (Haldol)	drowsiness, apathy, extra-pyramidal symptoms, dystonia, akathisia, hypotension, constipation, dry mouth, weight gain, tardive dyskinesia
		Risperidone (Risperdal)	sleep disturbance, agitation, anxiety, restlessness, headache
		Fluphenazine (Prolixin)	sedation, extra-pyramidal symptoms, dystonia, akathisia, hypotension, constipation, dry mouth, weight gain
Antidepressant	**Selective Serotonin Re-uptake Inhibitors (SSRI)**	Fluoxetine (Prozac) Sertraline (Zoloft) Paroxetine (Paxil) Citalopram (Celexa)	insomnia, diarrhoea, gastro-intestinal upset, restlessness, weight loss, dry mouth, anxiety, headache
	Tricyclics	Nortriptyline (Pamelor), Amitryptiline (Elavil)	insomnia, diarrhoea, gastro-intestinal upset, restlessness, weight loss, dry mouth, anxiety, headache
Antiepileptics		Valproate (Depakote) Topiramate (Topamax) Carbamazepine (Tegretol)	nausea, vomiting, weight gain or loss, cognitive effects, tremor, and elevated liver enzymes

[3] NB: Some drugs have dual actions. Not all drugs listed are available in all European countries

2. Role of the Physiotherapist and the Potential Benefits of Physiotherapy

Physiotherapy is recognized as a health care profession, which utilizes "physical approaches to promote, maintain and restore physical, psychological and social well-being" (Chartered Society of Physiotherapy 2002). The physiotherapist aims to promote quality of life and independence by encouraging activity and providing support within functional tasks (Royal Dutch Society for Physical Therapy, 2004). Physiotherapy is also focused on safety and interventions may be aimed at the prevention of falls (Royal Dutch Society for Physical Therapy, 2004).

The beneficial role of physiotherapy within basal ganglia disorders has been previously illustrated within Parkinson's disease (PD), with two recent systematic reviews reporting that physiotherapy can improve multiple factors including physical functioning, health-related quality of life (HR-QoL), strength, balance and gait (Goodwin et al. 2008; Kwakkel et al. 2007). The literature in support of physiotherapy for people with HD is less clear. Two reviews have noted that there is a small amount of evidence supporting physiotherapy within HD. This is somewhat overshadowed by poor methodological rigour, small sample sizes, and unclear selection criteria, resulting in potential heterogeneity in participant groups, and a lack of follow-up (Bilney et al. 2003b; Busse & Rosser 2007). There is however a growing body of recent literature supporting the use of physiotherapy in HD. For example, a before-after trial with a sample size of 40 found an intensive rehabilitation programme of 6 sessions per week demonstrated an improvement in motor function over the two year period (Zinzi et al. 2007). Furthermore, positive findings from environmental enrichment studies in mice provide some support for the basis of physiotherapy for people with HD. Mice with HD, placed within an environment providing physical, mental and social stimulation, have a slower disease progression, and maintain motor function for longer (Dobrossy & Dunnett 2005a; Hockly et al. 2002). Based on these studies and the biological rationale, there is a place for continuing to explore the use of physiotherapy within the management of people with HD. In depth efficacy studies are still required. In the interim, these guidance notes, which provide a written framework for the physiotherapeutic management of people with HD, hope to provide international uniformity of care that allows practitioners to make an informed decision regarding patient care.

3. Physiotherapy Evaluation

3.1 Overview

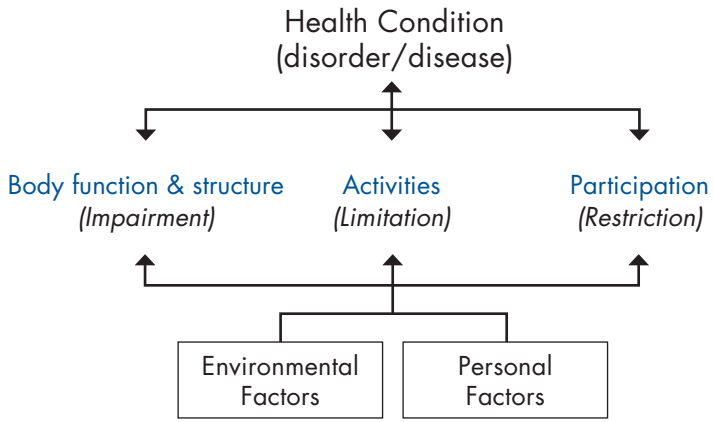
The important components of a physiotherapy evaluation for people with HD include descriptive evaluations (namely describing the nature of problems at the level of participation, activity, and body structure and function), and discriminative evaluations (i.e. distinguishing "normal" from "abnormal" function). The latter is particularly important for people in pre-symptomatic stages. Evaluation is also important in terms of providing baseline measurements for future comparisons (see Section 4: Outcome Measures for a discussion of relevant evaluative measures).

The EHDN-PWG recommends using the International Classification of Functioning, Disability and Health (ICF) (WHO 2001) to structure a patient evaluation (see Figure 3). We have incorporated terminology supported by the Guide to Physical Therapist Practice (American Physical Therapy Association 1999) to describe the evaluation process.

We provide an Evaluation Framework for conducting evaluations of patients with HD following the ICF framework. In the Evaluation (Section 3), we discuss a) Reason for Referral b) Social History and Participation Restrictions and c) Limitations in Functional Activities and d) Impairments in Body Structures and Functions. In Treatment Planning (Section 4), we refer to a) Assessment b) Goals and c) Plan of Care.

This evaluation framework for people with HD follows the ICF model, and is adapted by work from Quinn & Gordon (2003). This framework describes aspects of a person's health and health-related wellbeing in terms of Participation Restrictions, Limitations in Functional Activities, and Impairments in Body Structure and Function. The specifications of each component are described in further detail below.

Figure 3: International classification of Functioning, Disability and Health (ICF)



The ICF further takes into consideration the contextual factors (both environmental and personal), which impact upon and shape a person's life, and notes how these feed into the body function/ structure, activities and participation sections. The ICF can be utilised as a model for a clinical subjective and physical examination to ensure a holistic assessment takes place.

For additional reading on physiotherapy assessment procedures, please see Susan Edwards (2002) Neurological Physiotherapy: A Problem-solving Approach, 2nd edition, Elsevier; Darcy Ann Umphred (2007) Neurological Rehabilitation, 5th edition, Mosby; or Susan B. O'Sullivan, Thomas J. Schmitz (1994) Physical Rehabilitation: Assessment and Treatment, 3rd edition, F.A Davis.

3.2 Framework for conducting Physiotherapy Evaluations in HD

This framework is structured to include those areas that we recommend evaluating in most people with HD, and what should be documented in an evaluation report. We have also provided specific guidance for evaluation procedures not commonly used in general physiotherapy evaluation (e.g. evaluation of dystonia). Standardized outcome measures that may be useful for people with HD are described in the following section (3.3).

3.2.1 Reason for Referral

In this section, the therapist obtains information pertaining to the patient's diagnosis and medical history. This should include date of diagnosis, confirmation of diagnostic testing, and reasons for referral at this stage. This section also includes current medications and may include family history. Specific components can include:

- Patient information and demographics
- Current condition: concerns that led the patient to seek services of physiotherapist; current and prior therapeutic interventions; current stage of HD and age of onset
- Past medical history: prior hospitalizations, surgeries and pre-existing medical and other health-related conditions; family history (if pertinent to course of treatment)
- Medications: list any medications taken for HD symptoms as well as for other medical conditions

NB: In multi-disciplinary specialty HD clinics, it may be unnecessary to document all of this information, if it has previously been documented by other members of the management team and if the information is readily accessible to the physiotherapist.

3.2.2 Social History and Participation Restrictions

The background history allows the physiotherapist to:
- gain subjective information on the patient's presenting problem(s)
- place this information in the context of their past medical history and current condition
- document how their presenting problem(s) may affect their ability to live independently and within society
- address the patient holistically

This section of the assessment provides the opportunity to build a therapeutic, collaborative relationship, enabling the patient to voice their expectations of physiotherapy, whilst the physiotherapist ensures realistic goals are set based on these expectations. It must be noted that the patient's caregiver(s) should be involved in this part of the assessment, particularly if the patient has difficulties communicating through cognitive or physical impairment.

This section can specifically include:
1. Home environment - i.e. living environment and community characteristics; family and living situation; family and caregiver resources; devices and equipment (e.g. assistive, adaptive, orthotic)
2. Employment/work - i.e. current work situation and requirements of position
3. Health status - i.e. prior functional status in self care and home management, including Activities of Daily Living (ADLs) and Instrumental Activities of Daily Living (IADLs) and physical function (e.g. mobility, sleep patterns, restricted bed days)
4. Environmental, home, and work (job/school) barriers
5. Community, leisure and social activity participation
6. General health perception/quality of life
7. Psychological function (e.g. memory, reasoning ability, depression, anxiety)
8. Behavioural health risks (e.g. smoking, drug abuse)
9. Level of physical fitness

3.2.3 Limitations in Functional Activities

For each functional activity, therapists should evaluate and document the level of assistance or caregiver burden needed to complete the task, and also determine the patient's skill level (e.g. time to complete task, consistency of task performance, etc).

This section can include any of the following subheadings or skills, as appropriate:

1. Self-care and home management (including activities of daily living [bed mobility; dressing; self care; toileting; bathing; eating; cooking, preparing meals and instrumental activities of daily living])

2. Walking/gait assessment – People with HD begin to exhibit walking impairments from early on in the disease. In the early stages of the disease, walking is slow (bradykinetic) and patients begin to show a widened base of support (Rao et al. 2008) and decreased stride length (Grimbergen et al. 2008). They will often exhibit uneven and highly variable step and stride lengths, and dystonic posturing of the arms and legs is common. As the disease progresses, patients become slower and more unsteady, increasing their risk for falls (Busse et al. 2009).

   Common gait impairments include:
   - Wide base of support
   - Uneven step/stride lengths
   - Involuntary movements (chorea) of the legs
   - Lack of consistency in gait pattern
   - Dystonia (posturing) of upper extremities and trunk, such as:
     - ankle inversion
     - external rotation of shoulder and flexion of elbow
     - fisting
     - internal rotation and extension of shoulder and elbow extension
     - combined lateral flexion and extension of the trunk
   - Non-reciprocal arm movements
   - Increased time spent in double limb support
   - Impaired reaction to externally or internally produced perturbations
   - Difficulty ambulating in open environments

3. Falls Risk – Since patients with HD are at risk for falls (Busse et al 2009 ; Grimbergen et al. 2008), a Falls Risk Assessment (Busse et al. 2009; Rao et al. 2009) should be conducted for patients even from the early stages of the disease. A falls history should be taken, which would include frequency, location, and time of day that any falls have occurred (e.g. during the past 3 months). This type of assessment is inherently subjective and depends on the patient's and/or family members recall. During the course of an intervention, therapists can ask patients to keep a falls diary, to record any falls or near falls. As part of a Falls Risk Assessment, therapists should ideally evaluate the home environment, to determine any environmental factors that may put the patient at greater risk for falls (e.g., loose rugs, poor lighting, etc).

4. Other Functional Skills
    a. Sit to stand – Therapists should assess a patient's ability to perform sit to stand, and stand to sit, movements from a variety of heights and surfaces. People with HD tend to have difficulty with this task, frequently using their hands to come to stand for increased stability. Problems with stand to sit tend to be related to poor eccentric control, and patients tend to fall into a chair without controlling their descending movement.
    b. Bed mobility – Bed mobility and comfort in bed should be assessed. Patients may have difficulty moving themself easily around the bed, and safely coming to the edge of the bed and up to standing. Most patients do not have choreic movements when they are sleeping, however they still may be restless at night.
    c. Sitting ability – From early on in the disease, many patients have a tendency to sit with a posteriorly tilted pelvis and bear most of the weight on their sacrum or lumbar spine area. As the disease progresses, patients will need increasingly supportive chairs or wheelchairs, with appropriate support and padding to minimize injury if a high degree of chorea is present. The type of chair that the patient commonly sits in should be assessed for its support and comfort, and ease of transfers.

3.2.4 Impairments in body structure and function

A systems review should include neuromuscular, musculoskeletal (posture, range of motion, pain and muscle strength) and cardio-respiratory and cardio-vascular evaluation with documentation of any associated impact upon functional abilities.

3.2.4.1 Neuromuscular Evaluation

Evaluating neuromuscular impairments is important in order to help therapists understand what factors are contributing to a patient's functional problems. It is important to note that not all impairments directly relate to presenting activity limitations. For example, while most people with HD have chorea, it is typically not the primary impairment contributing to difficulties with ambulation. Bradykinesia and balance impairments may be better predictors of ambulation difficulties and falls (Busse et al 2009).

Dystonia is defined as abnormal, sustained posturing of any part of the body. A therapist can observe dystonia when a patient is sitting or standing (Louis et al. 1999) and in movements such as shoulder elevation, foot inversion and supination, and trunk extension. Some of the most common forms of dystonia seen in patients with HD are internal shoulder rotation, sustained fist clenching (47.1%), excessive knee flexion, and foot inversion.

Chorea is defined as involuntary, writhing movement. In the early stages of HD, it is typically seen in the fingers, hands and face muscles. As the disease progresses, it can be seen throughout the body, including in all four extremities and the trunk. The severity of chorea (maximal chorea) can be rated on the UHDRS Motor Rating Scale (score 0-4) where 0 is absent; 1 is slight/intermittent; 2 mild/common or moderate/intermittent; 3 is moderate/common; and 4 is marked/prolonged.

Balance problems typically occur in the early-mid stages of the disease, which interfere with a patient's ability to walk and maintain an upright position. Balance can be assessed by measuring proactive (e.g. reaching ability) and reactive balance (e.g. response to external perturbations). Scales such as the Berg Balance Scale (Berg et al. 1992a) or the Tinetti

Balance Assessment (Tinetti 1986) are useful in quantifying the degree of balance impairment and providing some insight into the nature of the balance impairments. Common conditions under which patients demonstrate balance problems include decreased base of support; tandem standing and walking; dual tasks; eyes closed and response to external perturbations (i.e. retropulsion pull test) (Bilney et al. 2003b; Quinn & Rao 2002).

Vision: People with HD often have impairment of control of the extraocular musculature; in particular they have difficulty with saccadic eye movements and with smooth pursuit (Hicks et al. 2008). This can frequently be seen in the very early stages of the disease. The resulting visual field impairments can have a significant impact on balance and walking ability, and thus therapists should have an understanding of the nature of a patient's problems. It may be useful for therapists to obtain this information from the UHDRS Motor section, which assesses both horizontal and vertical saccades and smooth pursuit.

Bradykinesia is defined as slowness of movement. Even though people with HD appear to be in constant movement, their underlying volitional movements during task performance have been found to be slower than healthy controls for reaching (Quinn et al. 2001) and ambulation (Delval et al. 2006; Delval et al. 2007). Bradykinesia can typically be evaluated by measuring time to complete a task (movement time).

Akinesia is defined as delayed initiation of movement, and in research studies is typically measured by reaction time. In the clinic setting, it is often difficult to quantitatively evaluate akinesia, but delays in onset of movement for various tasks can be noted.

3.2.4.2 Musculoskeletal evaluation

Posture and range of motion: There should be a standard assessment of posture and range of motion (ROM) in sitting and standing, and in supine for patients in all stages. The impact of posture and ROM on function, specifically positioning during performance of ADLs, should be considered.

Dystonia will have an effect on a patient's posture and ultimately on their range of motion if the dystonia is persistent. Patients with a high degree of dystonia should be carefully evaluated for range of motion limitations and muscle shortening.

One of the most common posture issues found in patients with HD is that of sliding down in a chair. Patients tend to adopt a posture of excessive thoracic kyphosis and posterior pelvic tilt in sitting, possibly as a strategy to increase their stability when their postural control becomes impaired, and as chorea becomes more prominent. By the later stages of the disease, patients will frequently adopt a more massed flexion posturing. This results in difficulty with upright sitting for eating, and also difficulty maintaining a standing position.

Strength: Individuals with HD have been found to have lower limb muscular weakness compared to age-matched controls (Busse et al. 2008a). It is unknown if this is a primary or secondary impairment, but regardless it can have a significant impact on a patient's functioning. Strength can be clinically measured by manual muscle testing or by functional observation of strength during task performance (e.g. stair climbing or evidence of gait deviation during walking).

Pain: People with HD can experience pain (Chudler & Wong 1995). The source of this pain can be unknown; however dystonia or mucle imbalances can often cause musculoskeletal pain. Excessive chorea can cause pain if people injure themselves by hitting their arm or leg into an object or hard surface. Some people may have pain unrelated to HD, such as a previous low back condition. When possible, therapists should ask people to rate their pain on a scale of 0-10 (0 being no pain; 10 being excruciating pain), and to describe the location and type of pain they are experiencing. For patients in the later stages of the disease who may have trouble understanding a 0-10 scale, a pain scale utilizing facial expressions, such as the Wong-Baker FACES pain rating scale can be useful (Wong & Baker 1988).

Figure 4: Wong-Baker FACES pain rating scale can be used for patients in the later stages of HD for communicating whether or not they are experiencing pain.

3.2.4.3 Cardio-respiratory and cardio-vascular evaluation

Regular monitoring of respiratory function in people with HD may be necessary. Lung function tests may highlight obstructive or restrictive disorders of the respiratory system. Measurement of vital capacity in supine and upright positions can identify weakness of the diaphragm (American Thoracic Society 2002). Forced expiratory Volume in 1 second (FEV1), Forced Vital Capacity (FVC), FEV1/FVC ratio, Peak Expiratory Flow Rate (PEFR) should also be considered using standardised spirometry techniques (Miller et al. 2005).

While dyspnoea is not a common symptom in people with neurological disease, it may be useful to monitor especially since it may indicate aspiration pneumonia, a common event in this disease. Two such scales for grading the degree of a patient's dyspnoea are the MRC Dyspnoea Scale, and the modified Borg Dyspnoea Scale (see Appendix 2).

The Medical Research Council (MRC) dyspnoea scale can be used to grade the degree of a patient's breathlessness. If a patient has a current respiratory problem, a specific respiratory assessment, including dyspnoea, should be undertaken using a modified Borg scale, which may be more sensitive to change over the period of the episode than the MRC Scale. Effectiveness of cough, whether the patient is able to clear secretions independently, and problems with swallowing should also be considered. Breathing pattern, lung sounds (auscultation) and exercise capacity (see Section 4.5) should be assessed objectively. Functional ability related to cardiovascular endurance should also be assessed and in particular whether there have been any changes in what the patient can do functionally. The 6-minute walk test (Enright 2003), which has been used in patients with cardiovascular conditions, as well as in the elderly and other patient groups (Falvo & Earhart 2009), can be a useful standardized assessment of general walking ability and cardiovascular endurance.

3.3 Outcome Measures

3.3.1 Overview

A physiotherapy outcome measure is a test or scale that accurately and reliably measures a particular patient's attribute, and which has the ability to be influenced by physical therapy intervention (Mayo et al. 1993). Thus, physical therapy outcome measures must be sufficiently sensitive to demonstrate change in a given domain over time. This is challenging for patients with degenerative diseases, such as HD, because simply having no change or deterioration in function may be a desirable goal. However, the use of standardized outcome measures for people with HD can be useful, and therapists should try to use one or more of these measures in an attempt to quantify changes in patient's level of participation, functional activities and impairments as a result of physiotherapy intervention. It is through the consistent use of such measures that therapists can begin to justify and support the benefits of physiotherapy for patients with HD.

3.3.2 Choosing outcome measures

There are a wide range of choices of outcome measures in physiotherapy; however none that are specifically for patient with HD. For more detailed information on using outcome measures, readers are referred to: *Fawcett AL (2007) Principles of assessment and outcome measurement for Occupational therapists and Physiotherapists. John Wiley & Sons: West Sussex, England.*

When choosing an outcome measure, there are several important considerations. First, therapists should determine what aspect they are attempting to measure. It may be useful to consider measurement tools based on the ICF model (Participation, Activity, Impairments in Body Structures and Function). Are the therapist and patient concerned primarily with walking ability or speed? Or rather are they both more concerned with participation in terms of functioning at work, or lessening caregiver burden?

Therapists should also choose measures that are reliable and valid. To date, very little research has been done to validate the use of outcomes measures in patients with HD. In the meantime, it is suggested that therapists utilize research from other neurodegenerative disorders (e.g. Parkinson's disease), to aide them in choosing appropriate tests. Definitions for commonly used terminology related to outcome measures are provided below. In the next section, we provide a list of potential outcome measures that can be utilized for patients with HD.

3.3.3 Outcomes measure terminology

Reliability – Reliability is a measure of how uniformly the test can be repeated when administered on more than one occasion (test-retest) or by more than one rater (inter-rater). Reliability of a test is typically measured by Intraclass Correlation Coefficients (ICC). Given that ICC values are susceptible to variation with increasing range in values, an alternative is to examine variability across test and re-test or across testers by computing the coefficient of variation (CoV). In addition,it may also be useful to conduct an analysis of differences between the two (or more) measures to evaluate if there are significant differences (using a t-test or ANOVA).

Validity – The validity of a test determines the extent to which a test measures what it is designed to measure. There are different types of validity, such as predictive validity (does the measurement predict a certain outcome), and criterion-validity (does the measure correlate with other well established measures designed to evaluate the same construct).

Responsiveness – Responsiveness is the ability of a test or measure to capture change over time. Is the measure sensitive to subtle changes related to an intervention?

Minimal detectable change (MDC) – MDC is the amount of change that must occur in a score to show that the change is not just due to measurement error. It is typically calculated using the standard error of measurement (SEM). The SEM represents how much a variable can vary within the subject, and is the standard deviation of the scores of a single person who is tested multiple times.

3.3.4. Relevant Outcome Measures

The following are a list of tests and measures that are recommended as potential outcome measures to be used by physiotherapists for patients with HD. We have categorized these tests and measures based on ICF level: Participation, Activities and Impairments (in body structure and function). When available, we have provided reliability (test-retest) and minimal detectable change for healthy adults, or for patients with PD or other neurological disorders, since no data is presently available for patients with HD. Versions of these measures can be found in Appendix 2, except where otherwise indicated.

3.3.4.1 Restrictions in Participation

Measures of participation restrictions can include evaluation of the following caregiver burden, ability to function within society, ability to care for oneself, as well as quality of life.

The Short Form-36 (Ware & Sherbourne 1992)
The SF-36 is a patient questionnaire that measures quality of life and level of participation. It is comprised of 36 questions that address 8 subscales. The SF-36 can not be reprinted, but can be found on the EHDN website (if you are an EHDN member; www.euro-hd.net Registry>Documents>CRF Forms)

Reliability – ICC >.80 (except for social functioning subscale which was .71; in patients with PD)(Steffen & Seney 2008)
Minimal Detectable Change – 19%-45% (in patients with PD) (Steffen & Seney 2008)

The WHOQoL Bref (Skevington et al. 2004)
The WHOQOL-BREF instrument was designed by the World Health Organization to measure quality of life across the following broad domains: physical health, psychological health, social relationships, and environment. It is comprised of 26 items, which ask the individual to score how they feel about their health and well being on a 5 point ordinal scale ranging from 1 – not at all to 5 - extremely. It has been validated in numerous countries and is available in many languages.

Reliability – ICC = .919 in elderly with depression (Naumann & Byrne 2004).

3.3.4.2 Limitations in Functional Activities

Six-minute walk test (Enright 2003)
This test requires a patient to walk around the perimeter of a set circuit for a total 6 minutes. Assistive devices can be used. Patients are timed, and the distance walked in 6 minutes is recorded. A patient's pulse can be taken before and after completion of the walk.

Reliability – ICC =.96 (in patients with PD) (Steffen & Seney 2008)
Minimal detectable change - 82 m (in patients with PD) (Steffen & Seney 2008)

10 m walk (Watson 2002)
A 14m walking area is marked, with 2 metres on either side allowed for starting and stopping of walking (acceleration and deceleration). Patients are asked to walk the length of the area at a comfortable walking speed. Patients can only participate in this test if they are able to ambulate without physical assistance, however assistive devices can be used. The total time to complete 10m, as well as the number of steps taken during those 10m are recorded. Average gait speed (distance/time) and cadence (step/min) can then be calculated. This test should be repeated twice and the average score for both tests documented.

Reliability – ICC = .87 (in patients with PD) (Schenkman et al. 1997)

The Timed Up & Go (TUG) Test (Podsiadlo & Richardson 1991)
For this test, patients stand up from a chair, walk 3 meters, turn around, walk back and sit on the chair. The test is timed. Patients should not participate in this test if they are unable to ambulate without physical assistance, but assistive devices can be used. This test is repeated twice and the average score for both tests documented.

Reliability – ICC .85 (in patients with PD) (Steffen and Seney 2008)
Minimal detectable change – 11 sec (in patients with PD) (Steffen & Seney 2008)

Physical Performance Test (PPT) (Reuben & Siu 1990)
The PPT is a compilation of items mimicking basic and complex ADL tasks and is scored by timing the completion of a task. This time then is related to a categorical score of 0 to 4, in which 4 represents people in the fastest 20% at completing the task, 1 represents those in the slowest 20%, and 0 represents those who cannot complete the task. The maximum score on the

9-item PPT is 36. Subjects perform a series of 9 standardized tasks, which are timed. The tasks include writing a sentence, simulated eating, turning 360 degrees, putting on and removing a jacket, lifting a book and putting it on a shelf, picking up a penny from the floor, a 50-foot walk test, and climbing stairs (scored as two items: time for climbing one flight of stairs and counting the number of flights of stairs the subject is able to ascend). Reliability and minimal detectable change do not appear to be reported.

Rivermead Mobility Index (Collen et al. 1991)

The RMI is a simple and short outcome measure, consisting of 14 questions and 1 observation. Its items cover a wide range of activities, from turning over in bed to running. The items are scored either unable or able (0–1) and added to produce a total score (0–15). A higher score reflects better mobility.

Reliability – .96 (in patients post stroke)
(Chen & Hsieh 2007; Liaw et al. 2008)
Minimal detectable change – 2 points (in patients post stroke)
(van de Port et al. 2006)

Barthel Index (Mahoney & Barthel 1965) (Hsueh, I. et al. 2002)

The Barthel Index is a 10 category rating scale that evaluates the level of assistance needed by a patient to perform the following tasks: feeding, bathing, grooming, dressing, bowels, bladder, toilet use, transfer, mobility and stairs.

Reliability – ICC >.8 (in patients post stroke)
(Hsueh et al. 2002)
Minimal detectable change – 1.85 (in patients post stroke)
(Hsieh et al. 2007)

Activities of Balance Confidence (ABC) Scale (Powell and Myers 1995)

The ABC is a questionnaire designed to measure a person' confidence in performing a variety of tasks without losing balance or becoming unstable. The underlying construct being measured is based on self efficacy theory (Tinetti et al. 1990)

Reliability – ICC = .94 (in patients with PD)
(Steffen and Seney 2008)
Minimal detectable change – 13% (in patients with PD)
(Steffen and Seney 2008)

UHDRS Functional Status (Huntington Study Group, 1996)

The UHDRS is a rating system used to quantify the severity of Huntington's Disease. It was developed as a clinical rating scale to assess four domains of clinical performance and capacity in individuals with HD: motor function, cognitive function, behavioural abnormalities, and functional capacity. These scores can be calculated by summing the various questions of each section. The functional capacity domain is a self-report measure of performance in various ADL's and IADL's. Research on the validity of the UHDRS indicates that it may be useful for tracking changes in the clinical features of HD over time, and appears to be appropriate for repeated administration during clinical studies. The Functional Status section may not have sufficient specificity or sensitivity for changes related to physical therapy interventions, but this has not yet been systematically evaluated.

Reliability – not reported

3.3.4.3 Measures of Body Functions

UHDRS motor (Huntington Study Group, 1996)

The UHDRS is a rating system used to quantify the severity of HD. It was developed as a clinical rating scale to assess four domains of clinical performance and capacity in individuals with HD: motor function, cognitive function, behavioral abnormalities, and functional capacity. These scores can be calculated by summing the various questions of each section. In the motor function domain, some sections (such as chorea and dystonia) require grading each extremity, face, bucco-oral-lingual, and trunk separately. Research on the validity of the UHDRS indicates that it may be useful for tracking changes in the clinical features of HD over time, and appears to be appropriate for repeated administration during clinical studies. The motor section may not have sufficient specificity or sensitivity for changes related to physical therapy interventions, but this has not yet been systematically evaluated.

Reliability – ICC was 0.94 for the total motor score, 0.82 for the chorea score, and 0.62 for the dystonia score (in patients with HD) (Huntington Study Group, 1996)

The UHDRS can be found on the EHDN website (if you are an EHDN member; https://www.euro-hd.net/html/registry/docs/forms)

Berg Balance scale (Berg et al. 1992)
This scale entails 14 sub-tests of various activities related to balance control. Sub-tests include static postures (e.g. sitting, standing), transitions (e.g. sitting to standing, transferring between chairs), and challenging positions (e.g. standing with eyes closed). Quality of performance is scored on a five-point scale. This test takes 15 – 20 minutes to administer.

Reliability – ICC >.90 (Parkinson's Disease)
(Steffen and Seney 2008)
Minimal detectable change – 5 pts (Parkinson's Disease)
(Steffen and Seney 2008)

Tinetti Balance and Gait (Tinetti 1986)
The Tinetti Gait and Balance Instrument was initially designed to determine an elderly person's risk of falling. It takes about 5 minutes to complete. It is a 2-part evaluation that measures performance on various balance tasks and provides a quantitative ranking of various gait deviations.

Reliability – ICC>.80 (in patients with PD) (Kegelmeyer et al. 2007)

Dynamic Gait Index (Shumway-Cook et al., 1997)
The Dynamic Gait Index was designed to evaluate a person's ability to modify gait in response to changing task demands. It is indicated for use with ambulatory patients with balance impairments.

Reliability – ICC = .96 (in patients with chronic stroke)
(Jonsdottir and Cattaneo 2007)
Reliability – ICC = .983 (in subjects with MS)
(McConvey & Bennett, 2005)

4. Physiotherapy Management/treatment

4.1 Goal setting

Goal setting is a critical component of any physiotherapy program for people with HD. The following are general guidelines for setting goals for people with HD:

- Physiotherapy goals should be functional, and should address specific functional problems or participation restrictions that are amenable to physiotherapy intervention.
- Goals should be focused on outcomes that are agreed upon by patient and therapist, and should be measurable and testable.
- Even though HD is a degenerative disease, it is still possible for physiotherapists to set goals such that functional performance will improve.

EXAMPLE 1:
A patient with **mid-stage HD** may come to see a physiotherapist because they have experienced falls, or are having difficulty walking (e.g. outdoors; in crowded environments, limited endurance).

Possible Goals:
1) Pt. will be able to walk outdoors for up to 10 minutes (avg. gait speed 1.1 m/sec), with HR <100 bpm, within 6 weeks.
2) Pt. will walk within home with 4-wheeled walker, avoiding obstacles and without falls within 4 weeks.

EXAMPLE 2:
A patient with **late-stage HD** may have difficulty maintaining an upright sitting position for eating and watching television.

Possible goals:
1) Pt. will maintain upright sitting for 20 minutes during eating while in adaptive seating system, within 2 weeks.
2) Pt. will sit comfortably in upright armchair with adaptive seat cushion for 30 minutes while watching TV, without sliding down in chair, within 3 weeks.

Goals to decrease chorea or dystonia are not realistic for physiotherapy. Furthermore, amelioration of any particular impairment in HD may not translate into functional improvements, and therefore therapists should focus on functional gains while attempting to ascertain the influence various impairments have on activitiy limitations. While it is reasonable that such impairments be addressed during physiotherapy intervention, the focus of goal setting should be on specific improvements in functional activity performance.

4.2 The Physiotherapy Assessment

Assessment is the process of analyzing the result from the physiotherapy evaluation. In the assessment section of an evaluation report, the therapist writes a summary paragraph that includes the following:

- Summary statement of the patient and the diagnosis
- Statement describing patient's activity limitations, key impairments contributing to those limitations and participation restrictions or potential participation restrictions that will result from those limitations
- Statement summarizing the patient's potential to benefit from physiotherapy and the reasons why physiotherapy treatment is or is not indicated

4.3 Plan of Care

Developing and documenting a Plan of Care for a patient with HD should include the following:

- Coordination/communication- The therapist should describe all coordination or communication that needs to take place with other professionals, caregivers, or family members so that the physiotherapy plan can be implemented. This can also include referrals to other professionals.
- Patient-related instruction - This should include any instructions given to the patient or their caregiver, and any planned home program and patient/caregiver education.
- Procedural interventions – This section should describe the specific physiotherapy procedures that will be conducted (e.g., gait training, balance training, ROM exercises, etc).

4.4 A framework for physiotherapy management of people with HD

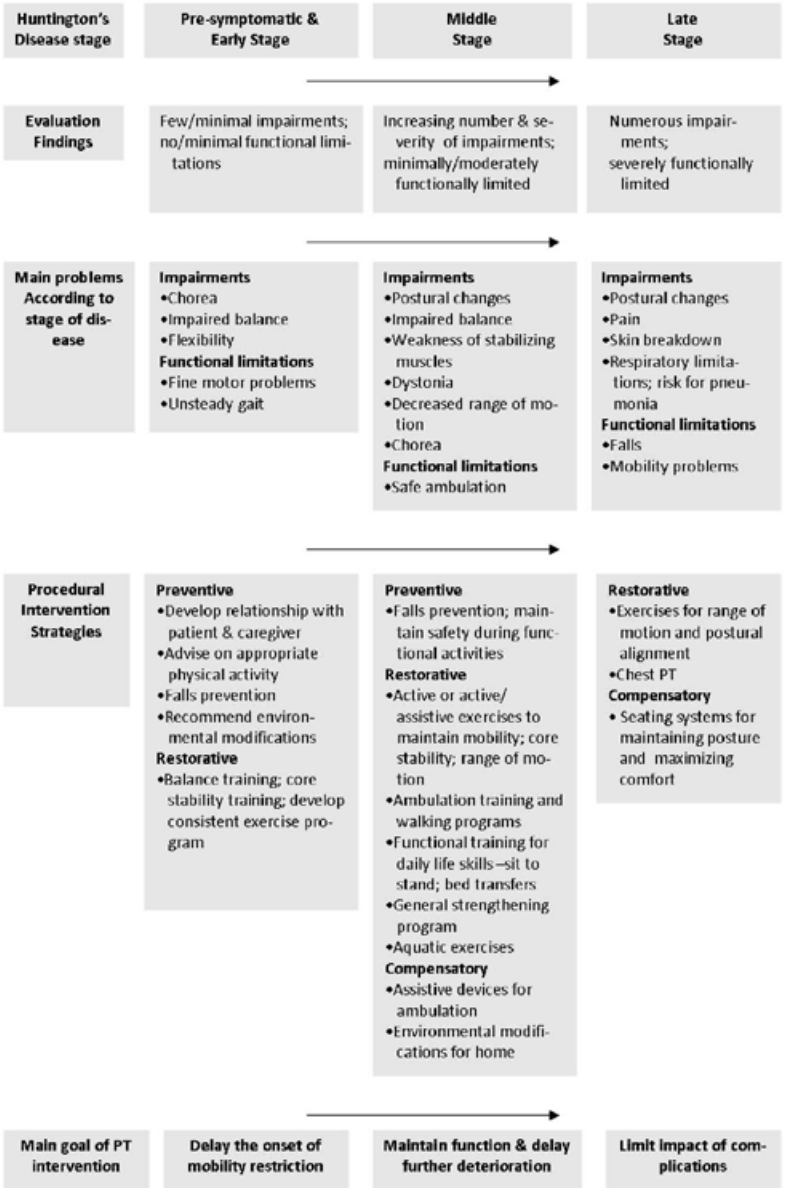
In terms of a general strategy for physiotherapy interventions, it is important to consider people at all stages of the condition, including those who have the mutation for HD but are not displaying motor symptoms (pre-manifest). It is further suggested that physiotherapy management of people with HD should be modified according to individual problems and to the stage of the disease. The framework presented in Figure 5 provides an outline for intervention according to the stage of the condition and potential impairments (Busse et al. 2008b). Importantly, physiotherapy should be part of an interdisciplinary team that is involved in the management of patients with HD throughout the lifespan of their disease. Such professionals include geneticists, neurologists, nurses, occupational therapists, psychologists, speech language therapists, and social workers, among others. It is critical for therapists to consider collaboration with these professionals and to understand their role as part of an interdisciplinary team.

4.4.1 Early referral to physiotherapy

It is recommended that referral occur early in the disease (or even in the pre-manifest stage), a view supported by physiotherapists practising within HD (Busse et al. 2008b), and within other basal ganglia disorders such as PD (Ashburn et al. 2004). There is increasing support for early intervention for people with HD, where an impact may be made on biological processes and potentially influence the natural history of the condition (see Figure 1, page 9).

Early referral to physiotherapy for people with HD may be beneficial to people in a number of ways. It enables practitioners to ascertain a baseline for the patient, supports the establishment of a therapeutic relationship between the patient, practitioner and caregivers, and ensures early intervention to maintain patient mobility and function for as long as possible (Busse et al. 2008b). It is important to note that in the early stages, patients may just be coming to terms with their diagnosis, and might not be willing or ready to accept advice or recommendations, and may have particular difficulty discussing what the future holds. The physiotherapist must be very sensitive to the patient's emotional state, which sometimes may necessitate progressing more slowly with an intervention plan than might ideally be indicated.

Figure 5: Conceptual Framework for Physiotherapy Intervention
(Busse et al. 2008)



4.4.2 Specific problems and potential treatment strategies

The main goals of physiotherapy intervention will generally change over the course of a disease; procedural interventions will initially be preventative, and gradually may become restorative (see Figure 5). Towards the end stage, physiotherapy interventions will be compensatory and will aim to limit the impact of deterioration and prevent associated complications.

It is important for physiotherapists to consider the cost implications of their services throughout a life-long disease. It is unreasonable to expect that any funding agency (government or otherwise) would be agreeable to ongoing physiotherapy intervention 2-3 times/week for the extended disease process. Instead, therapists should consider serving consultative roles to patients from an early stage, and providing more intensive intervention when a change in functional status warrants it.

When determining potential treatment strategies, it is important first to identify the problem and its cause. For example, involuntary movements could contribute to balance problems, but inactivity may also be a contributing factor. Accurate assessment is therefore essential to the development of any treatment programme. Impairment-based diagnoses that may impact on function include: dystonia; bradykinesia; moderate-severe chorea or ballismus; rigidity; impaired respiratory function and fatigue. It is important to manage the secondary effects of such impairments. For example, for patients with chorea protective equipment can be provided, and for patients with dystonia, loss of range of motion and muscle imbalance should be prevented. Functional problems that may occur include: impaired fine motor skills and manual dexterity, impaired sitting posture and sitting ability, impaired mobility, transfers and gait; impaired balance/ and risk of falls and reduced cardiovascular and general physical fitness. In general, it is thought that physical fitness plays a part in (i) a patient's motivation to keep as active as possible and (ii) their ability to cope with more challenging treatment interventions. Those patients who have always led an active lifestyle prior to onset tend to do better, especially in the early stages. The potential impact of psychiatric impairments and a patient's cognitive status cannot be overlooked. Therapists should consider whether or not a patient has memory loss, depression, aggression, obsessive-compulsive tendencies, or anxiety, to name a few, and these impairments must be taken into account during the design of any intervention plan.

4.4.3 Procedural interventions

In this section, we describe commonly utilized intervention strategies for people with HD. When available, we cite supporting research related to these interventions. However in many cases, the recommended interventions are based on expert opinion, and should be re-considered as new literature becomes available.

4.4.3.1 Balance, co-ordination and core stability training

Interventions for retraining balance in individuals with HD depend on the underlying neural, motor and sensory impairments that contribute to postural instability and balance problems. Postural control impairments are closely linked to gait abnormalities, thus interventions for gait problems may indirectly or directly affect balance. In general, balance interventions should focus on: (1) restoring underlying impairments whenever possible; (2) preventing secondary impairments that may affect postural instability; (3) facilitating task-specific sensory and motor strategies necessary for meeting postural control demands; and, (4) practicing the maintenance of postural control in a variety of tasks and environments. Balance training activities, in general should be task specific (Shumway-Cook & Woollacott 2006). Progression from a wide to narrower base of support, from static to dynamic activities, from a low to high centre of gravity, and increasing the degrees of freedom that must be controlled can be considered. The key principle is that the balance demand of a specific task should be assessed and addressed.

In addition, an important component of physiotherapy even from the earlier stages is teaching people to get up from the floor. This is particularly important if patients are being given a home exercise program that requires them to get down onto the floor. Even those patients who have become cognitively impaired may be able to copy a demonstration of how to do it and repeat it until it becomes an automatic motion, even if they can't cope with more complex concepts such as backward chaining (Reed & Simpson 1996).

When restoration is not possible, compensatory interventions may be necessary to address sensory, visual, vestibular and motor impairments, in order to promote balance safety (Busse et al. 2008b). A careful examination should be conducted of the home, work and community environment in which the patient must function, and modifications made accordingly. In addition, prescription of assistive devices, home adaptive equipment or modifications may be required. Balance training and strengthening of the postural muscles (core stability) to enhance postural stability should begin in the early stage of the disease and should take place in the environment where the individual's problems are most apparent, if possible.

People should be taught to deliberately prepare in advance for forthcoming threats to balance, or to focus their attention on maintaining balance before a task in which equilibrium is challenged is initiated. It is thought this strategy allows people to use frontal cortical systems to regulate stability, rather than rely upon the impaired basal ganglia mechanisms (Bilney et al. 2003a). Training the patient to step in response to perturbations, with an emphasis on speed and accuracy of the stepping strategy, is also recommended.

To address the patient's inability to multi-task at a time, interventions could include instructing the patient to attend to one task at a time (Delval et al. 2008b). People at high risk of falls should be taught to break down complex activities into simple tasks and to focus their attention on performing each task separately; alternatively, having people with HD practice doing two activities at the same time under various practice and context conditions may be a beneficial component to balance training (Delval et al. 2008b). In addition, doing exercises that requires automatic responses, i.e. throwing a ball, may be of benefit in eliciting postural responses and training to move more quickly. Fatigue may also contribute to falls risk; strategies should be provided for the person with HD to help them identify when fatigue and the associated influence on performance of functional activities could increase their risk of falls.

4.4.3.2 Functional and task-specific training

Functional training is a critical component of any intervention plan for people with HD. This consists of assisting people with HD with strategies to directly improve their ability to perform daily functional tasks, such as dressing, bathing, climbing stairs, etc. Functional training also includes device and equipment use and injury prevention or reduction (Busse et al. 2008; Quinn & DalBello-Haas 2005; Quinn & Rao 2002). It is also useful to find out any hobbies or interests that the patient is interested in now, or was before the onset of the disease, and use of such activities can assist in their rehabilitation program.

Task-specific training is particularly appropriate for people with movement disorders (Bilney et al. 2003a), because motor disturbance is typically context-dependent, and is typically seen in complex, well-learned tasks such as walking and reaching. Motor learning research suggests that task-specific practice is essential for permanent improvement in functional abilities (Berhman et al. 2000; Dobrossy & Dunnett 2005b). A task-based model of intervention is designed to enhance skill learning, and is particularly important for people with degenerative neurological diseases. A task-based model involves first the determination of the task to be learned and setting an appropriate goal for skill attainment (Berhman et al. 2000; Gentile 2000). This will typically involve re-learning a previously learned skill, such as walking, but while using adaptive equipment. A person's level of skill performance and ability to learn a new skill (either explicitly or implicitly) should be considered in designing an appropriate task-based intervention, which involves structuring practice sessions to maximize learning, as well as structuring the environment, and providing augmented information before, during and after task performance.

Because cognitive impairments also increase in number and severity as the disease progresses, additional compensatory strategies, such as providing cues with goal directed feedback, teaching skills using one step commands, and providing treatment in a quiet, non-distracting environment should be incorporated. Practice should take place through repetition, allowing sufficient time for the person with HD to understand what is required of them and with key points continually being reinforced (Quinn & Dal Bello-Haas, 2005).

4.4.3.3 Ambulation and Gait Training

The primary goal of people with HD, related to physiotherapy, is often to improve walking ability (speed, coordination, balance, distance, or to simply continue walking for as long as possible). The focus of gait training should be on identifying those aspects of gait which are functionally limiting (Rao et al. 2008), and then designing an intervention plan that is aimed at ameliorating or compensating for the gait impairments, and providing training to people with HD so that they can reach their ambulation goals. The influence of the practice environment on learning and carry over and the interaction between the patient and the environment directly affects the movement that emerges (Bassile & Bock 1995). Structuring practice of walking within an environment that is realistic to the patient's current life situation is likely to yield the best results.

As the disease progresses, compensatory strategies such as using sensory cues or attentional strategies, can be implemented. Assistive devices and safety equipment, such as a helmet or elbow and knee protectors, may be recommended (see section on Assistive Devices below, and also Appendix 3). In some therapists' experiences, one-person assistance can be safer than use of an assistive device. Having a caregiver walk alongside the patient, holding onto the patient's hand or waist, or walking behind a patient and providing support at the pelvis allows the caregiver to more easily adjust to the patient's altering movements, so that the walking becomes both smoother and safer. Some patients who have even older spouses/ carers with their own mobility or health problems may find this difficult and may be unable to safely provide this assistance.

Walking aids

Currently there is little research evidence of the most suitable equipment for people with HD. Walking and balance impairments in people with HD can result from subtle changes in co-ordination, involuntary movements and prolonged muscle contractions, and the presence of dystonia (abnormal posturing). These changes can lead to problems in walking, in and out of the home and over long distances, or increase the number of falls or accidents, which can lead to fractures or other bodily injuries. In the early stages of HD walking aids are usually not required, however as the condition progresses to the middle stages walking aids are more relevant.

A rollator walker with swivel castors is a commonly recommended device for individuals who are having difficulty walking unaided. A wheeled walker seems to provide many patients with good stability, and has been found to produce the most typical gait pattern when used by people with HD (Kloos et al. 2009). Those patients who can cope with the brakes can use them to slow their forward motion. Walking sticks tend to be more hazardous for patients with HD, particularly if they have chorea of the upper limbs. Weighting down of a walking stick (by inserting sand or coins into a hollow aluminium stick, or by wrapping a weight around the base of a wooden stick) may be helpful for some patients. People early in the disease process sometimes like the idea of some type of device so that it alerts people that they have difficulty with walking.

Specific considerations when recommending walking aids for people with HD include:

- Person's ability to adapt their walking with their involuntary movement patterns
- Fatigue
- Environment
- Hand function: the ability to hold handgrips on equipment
- Ability to operate brakes on wheeled frames
- Lack of concentration
- Memory: impairment/appropriate methods of facilitating (psychology guidance)
- Compliance issues, difficulty in accepting change.
- Aggression: risk to self/others/environment (may be variable and recognition of triggers and management important)

Table 2 contains details of some assistive devices (canes and walkers) which can be helpful to prevent falls and increase the amount of time and distance for walking, while allowing a person with HD to maintain as much independence as possible. Sometimes it is safer to have no aid or a fixed rail on the wall. When walking becomes very difficult for people with HD, a wheelchair may be prescribed. A wheelchair can be beneficial for daily use or for long distance mobility (see Seating Considerations section). People with HD can also choose to use the wheelchair for part of the day (start out the day by walking and then using the wheelchair as the day progress or when fatigue sets in).

Table 2: Walking Aids for consideration

Canes/sticks Function: to widen the base of support, improve balance but does not provide much stability	
Types	**Considerations**
Straight canes **Height adjustable canes**	• Supports only small % of body weight. A heavier stick helps with co-ordination. • Weight can be added to sticks which are hollow by removing the rubber infill and filling the base with coins or sand if sealed.
Quadrapod sticks: small/large base centered or asymmetrical	• A greater risk of tripping with a symmetrical/centered aid as 2 feet are adjacent to the walker's foot. • An asymmetrical quadrapod is less stable but there maybe less risk of tripping over its legs.

Types	**Considerations**
3- and 4-wheeled walking frames **Modifications to consider:** 1) Locking of the folding system so that it cannot be released whilst in use. 2) A heavier frame is easier to control, some are made of steel,weight can be added to the basket. 3) If it has a seat it is only safe with a back otherwise modify it so that either the seat has a tray or a back 4) Brakes can be modified, different systems	• 3-wheeled frames for some, but are less stable than 4-wheeled on turning and with more pressure on one side if one hand releases it may tip and person may fall. • Larger wheels move better over uneven surfaces indoors and out. • Applying brakes may be a problem for user who has to dual task due to memory problems and lack of insight if unsupervised. • Some people tend to lift their frames as they jerk backwards. • Can be cumbersome and difficult to manoeuvre through some areas.
4-wheeled forearm support frames **Function:** To give more support through the forearms , particularly if hand function is impaired. **Types:** 1) Gutter frames 2) Split level forearm support 3) Frame with single padded support.	• May be cumbersome. • May give an opportunity to maintain mobility for longer. • Supervision or assistance may be required. • Some hard surfaces under supports which may need padding to prevent self injury from involuntary movements.
Frames with 2 front wheels and 2 rear feet	• Useful when brakes can not be used and the frame has a tendency to run away. • Difficult to move over carpet pile.

Strategies to reduce the risk of injury when walking include:

- Use protective helmets/hats, elbow pads, knee pads and hip protectors.
- Create a limited, safe area to allow ambulation.
- Supervise, guide and prompt to promote safe ambulation.
- Provide physical assistance: hand holding; support though hips; use of gait belt.
- Train family/carers; re-enforce consistent technique of walking.
- Facilitate access to re-assessment/treatment as mobility/compliance change.
- Encourage use of appropriate footwear.

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. This foundation should be stable and should provide support 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. Because shoes wear down very easily, it is important to consider buying new shoes every 6 months.

The following is a list of what to look for when choosing footwear to promote function safely:

Recommendations for choosing dress/casual shoes:

- Firm, flat heel
- Forefoot should be bendable
- Velcro or elastic shoelaces for ease of application
- Wide heel base for increased stability
- No thick soles
- No thick toe grips (thick toe grips and soles can catch and lead to falls)

Recommendations for choosing trainer type shoes:

- High tops recommended for ankle support
- Sneakers should be leather for support of heel and ankle
- Velcro or elastic shoelaces for ease of application
- Wide heel base for increased stability
- No thick soles
- No thick toe grips (thick toe grips and soles can catch and lead to falls)

4.4.3.4 Sensory cueing

Sensory cueing involves the use of augmented sensory information, typically in the form of external visual, auditory or manual cueing, to improve performance in a task. Sensory cueing has been most commonly used for people with PD as a means to facilitate movement and overcome episodes of freezing and akinesia, typically during gait. A major aim for therapeutic intervention in HD is for therapists to teach people with HD how to effectively bypass damaged basal ganglia structures, and use frontal neural pathways to control movement (Bilney et al. 2003a). This involves accessing the motor system by the patient responding to a visual or auditory input (which occurs through frontal pathways), rather than self-initiating movement (which occurs through the basal ganglia-cortical pathway). There is some controversy as to whether individuals with HD can benefit from rhythmic auditory cues to improve gait characteristics (Delval et al. 2008a; Thaut et al. 1999). Synchronizing gait to rhythmic cues from a metronome (but not music) has been shown to assist the modulation of gait speed in people with HD (Thaut et al. 1999) however under dual task conditions metronome cues may be less helpful due to attentional deficits in people with HD (Delval et al. 2008a; Delval et al. 2008b).

4.4.3.5 Seating Considerations [4]

Although many people with HD can walk independently or with some assistance, most have at least some difficulty with sitting comfortably. Most people have difficulty sitting in a chair with inadequate back and side support. Many people with HD tend to slide down in their chairs, and often maintain weight bearing through their lumbo-sacral spine as opposed to through the ischial tuberosities, buttocks and thighs. Additionally, people with choreic movements are prone to injuries if their chair is not properly padded. Persons with HD, especially in the middle and later stages, spend a majority of their day sitting. It is important to maintain an upright position especially for feeding and drinking to minimize risk of aspiration. Effective positioning with the appropriate supports will also enable people with HD to better interact with their environment and have an improved quality of life. This is especially important in the late stages of the disease as people are likely to be wheelchair dependent.

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 (Quinn & DalBello-Haas, 2005). Use of certain devices and equipment, such as those described above, may provide the necessary support to maximize a person's functional abilities.

[4] References: Brendon Stubbs, senior physiotherapist, St. Andrews Healthcare, HDA Newsletter June 2007 and NZ Huntington News, ed.99. Dec. 2007
Disabled living Foundation: www.org.uk/factsheets Walking aids, Wheelchairs, Seating
Eastin: European Assistive Technology Information Network www.eastin.info
Collaboration of Denmark, Nederlands, Germany, Spain, Italy and UK

Considerations for optimal seating include:

1. Chair measured so that the correct depth, height and width for the individual
2. Allow enough room for person to move around freely and without injury
3. Protect from hard surfaces and sharp edges with proper padding
4. Maximize ease of transfers
5. Provide for independent mobility if appropriate
6. Solid, sturdy foot support
7. Appropriate height for use at table or with lap tray
8. Minimal use of restraints

Progression from independent ambulation to use of a wheelchair can be very traumatic for the person with HD. This is a sign of their continued functional decline and lack of control over their life. Recommending a wheelchair as a primary means of mobility should be approached cautiously by the therapist. This decision should be reached with the consent of the patient, the family and the interdisciplinary team. Allow the person with HD to make their own choices to the extent that their safety and the safety of others are maintained. When 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 3 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)	• The range of movement of hips, knees and ankles; also the length of the hamstrings need to be maintained to maintain/improve seatability. Wedge the seat: deep at the front graduated to the back to adjust the angle at the hip (thigh-trunk segment) in conjunction with a tilt in space chair. Where the angle of extension of the knee is less than 90 degrees and the high tone of the hamstrings is pulling the client's pelvis forwards, the angle at the hip needs to be opened. • Seat cushion: needs to have the appropriate rating for skincare and to provide adequate support. An ischial cut-out to anchor pelvis in place could be considered. Check seat length is not too long.
Poor postural stability	• 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	• Use a chair with the least amount of chrome or metal exposed. • 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. • 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 sides	• 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 such as foam carve to provide good postural support.
Unable to tolerate upright chair; falls out of standard chairs	• Use a maximally adjustable chair with recline and tilt. • Change positions frequently (e.g., 20 minutes upright, 20 minutes fully reclined). • Use pillows, padding to provide extra support/protection; • Consider the use of bean bag chair, hammock, or padded floor bed.

Table 4: Wheelchairs and seating systems for consideration

Standard Wheelchairs: • 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: • Padding for the armrests & footrests to prevent bruising. • Spoke guards to cover the wheel spokes.
Reclining Wheelchair: • 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: • 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 • 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.

4.4.3.7 Bed positioning

In the middle and late stages of Huntington's disease people may develop more problems with positioning in bed. Problems include falling out of bed due to choreic movements and restless sleeping. Possible solutions are use of a double bed, padded bed sides or a floor bed. Other problems may include sustained dystonic posturing leading to contractures especially at the hip and knee. Bed positioning devices such as padded rolls and supports may be useful.

4.4.3.8 Protective techniques

Protective techniques are often used for people with Huntington's disease, who have an increased risk of falling (Grimbergen et al. 2008). Most injuries as a result of a fall tend to be minor (Grimbergen et al. 2008), however bruising and skin breakdown is not uncommon. People with HD can benefit from protective padding on the elbow, forearms knees and shins to minimize injury. In order to maximize a patient's ambulatory independence, a therapist may recommend use of a helmet and other body padding as a patient with HD advances in the disease process. A helmet may prevent serious head injury in case of a fall, but not subdural haemorrhage. A helmet may allow a patient who might otherwise be considered "unsafe" the opportunity to continue to ambulate independently. Patients may still have a difficult time accepting any form of protective equipment, and therapists need to work within the patient's needs to determining what is necessary. Overall, restraint should be kept to a minimum. Patients with involuntary movements will often act adversely to being constrained in any way, and therapists should always work to find ways to keep them safe within their environment without the need for restraints.

4.4.3.9 Airway Clearance Techniques

Respiratory problems may develop in any disease stage, but are more likely to occur in the middle and late stages of the disease as mobility becomes more limited. Physios should work in conjunction with speech and language therapists to address swallowing and respiratory issues. Respiratory interventions, such as deep breathing exercises, body positioning to optimize ventilation-perfusion, modified postural drainage, and airway clearance techniques may be required for specific respiratory problems. Caregivers may be taught assisted coughing techniques and a suction machine should be made available for those people with HD who have difficulty clearing secretions. Educating caregivers regarding the signs of aspiration, and also how to perform the Heimlich manoeuvre, is also important.

4.4.3.10 Exercise for people with HD

Any person with HD who is considering participation in an exercise programme should have a fitness profile created that considers their medical presentation, and the factors discussed above in the assessment but focuses on functional activities they need or want to be able to achieve. These factors will drive the exact mode of prescribed exercises, specifically frequency, duration, intensity and mode of exercise. Frequency and duration will depend on the overall cardiovascular fitness of the individual and any associated undesirable effects. Some people with HD will find that shorter episodes of exercise more frequently throughout the week to be less fatiguing, for example. Mode of exercise depends on the ability of the patient, although in terms of the exercise profile, a functional activity exercise mode is the optimal approach. This focuses on training the specific skills that the person with HD wants to improve i.e. hand exercises, sit to stand exercises or walking exercises.

Mode specific training will drive specific changes. Balance skills can also be trained during strengthening or endurance activities in a gym by using free weights or pulleys (low resistance). Limitations of the condition may necessitate the use of specific or adapted equipment. Adaptations will have to be made that consider a patient's medical and social situation as well as personal preference relating to exercise. Adherence is likely to be better if the person with HD and their carer are involved in the choice of exercise mode and the exercise profile that is developed.

In terms of the dose of exercise, the components of exercise that need to be trained (muscle endurance, strength, speed power, cardiovascular and flexibility) need to be considered. The muscles and joints that are required to do the movement need to be specifically targeted and the length of time should be related to the requirements of the functional activity. For example, if training for walking to the shops then it is important to train for the appropriate walking time (endurance) as well as considering speed in terms of requirements in the community for walking and crossing roads. A warm-up and cool-down should be incorporated into all programmes and as in other populations, careful baseline testing should be carried out prior to prescription. See Appendix 4 for an example of a case study applying the above principles in a person with HD.

The minimal dose and dose response to exercise are as yet not known in HD. Current recommendations are to utilise the American College of Sports Medicine guidelines for exercise prescription. It is however essential to review the response and employ careful monitoring throughout as safety and efficacy trials have not been conducted. Vital signs, symptoms of exertion such as dyspnoea, excessive fatigue, pallor, and dizziness, and signs and symptoms specific to HD should be monitored and documented, at rest and during and after exercise. The Borg Rating of Perceived Exertion (RPE) Scale (Borg 1982) can be used to objectively record perceived exertion.

The current American College of Sports Medicine (2009) guidelines for exercise prescription are as follows:

1. **Healthy individuals should exercise 3 times a week**

2. **For cardiovascular health 30 minutes at each session is required. It is generally considered safe to do 2 x 10 repetition maximum sets for the muscle groups you wish to train.**

3. **Intensity of training should be in the aerobic training zone of 50-80% age predicted maximal heart rate and for strength reps 60% of 1 repetition maximum for 10-15 reps.**

It is apparent that exercise prescription for people with HD may have similarities with what is known in other neurological conditions. However HD is a progressive disease and this requires skilful progression of exercises within the limitations of that individual. This may mean at times increasing the exercise and at times reducing the absolute amount of activity or the absolute load that is lifted or the treadmill or bike speed or resistance, whilst maintaining the relative intensity of the exercise as well as its duration and frequency. In some cases, the mode of delivery may be completely changed where impairments such as cognitive, behavioural or motor issues prevent the current mode of delivery. In these cases novel approaches such as using computer simulated activities (for example Nintendo Wii Sports) should be considered.

Exercise prescription for an individual with HD is further complicated by reduced motivation. It is important to consider long-term delivery issues. This may mean altering the setting of intervention i.e. in a gym or clinic or home-based. There are several home-based videos which have been recently designed specifically for people with HD, and are currently in the process of being evaluated. Furthermore the structure of the intervention i.e. group based vs. individual should be considered. The importance of group exercise for their competitive elements as well as social interaction cannot be overlooked. Even patients who are non-ambulatory can participate in chair-based group exercise programs.

The issues of support for the carers and the importance of involving the caregiver/ family in exercise should to be highlighted. The impact of any cognitive impairment and fatigue on the timing of an intervention during the day and the content of the intervention i.e. amount of physical activity/

advice/education given should also be considered. In order to ensure safety, careful instruction of safe exercise may be required. Furthermore, many affected individuals develop an intractable apathy that is linked to decline in cognitive function. Utilizing carers and engaging the patient in meaningful and motivating discussions about the benefits of exercise and therapy is critical.

Horse riding and therapeutic riding is an activity that has benefits for balance and core strengthening. Many patients like horse riding at early and mid stages, and riding programs for people with early and mid-stage HD have been used in HD Camps in the U.S. and Canada for a number of years.

4.4.3.11 Other Procedural Interventions

Manual therapy, electrotherapeutic modalities and physical agents as well as mechanical modalities are not discussed in these guidelines. These interventions are not commonly utilized in people with Central Nervous System Disorders, and although in certain cases their use may be indicated, there is no evidence to support their use in people with HD.

Relaxation exercises are intended to help minimize chorea and to enhance lower abdominal breathing. Also manual techniques such as massage or tactile stimulation, can be very effective to lower the degree of chorea or dystonia temporarily, but not on a long term basis. But this is nonetheless sometimes very helpful and calming for the patient if one can minimize the chorea for that instant. Massage can be a useful intervention to aid relaxation. It is also readily accepted by patients as they can just enjoy it and it doesn't involve work for them. There is some anecdotal evidence to suggest that it results in behavioural improvements, so it may be worthwhile in some patients. Finally, hydrotherapy can be a very useful intervention for patients with HD (Sheaff et al 1990). The buoyancy of the water can aid in ease of movements, and chorea tends to be minimized. It may be a very important alternative intervention for patients who have difficulty with traditional exercise, and it further offers the benefit of social interaction in a group setting.

Appendix One:
Review of available literature relative to physiotherapy in HD

Databases searched:
Ovid MEDLINE(R) 1950 to March Week 2 2009
EMBASE 1980 to 2009 Week 12, 21st March 2009
EBSCO CINAHL 1981 to 21st March 2009
PEDro 1929 to 21st March 2009

Search Strategy:
Articles published in English; Therapy in human HD subjects only; Subjects to be of 18 years of age + with a confirmed diagnosis of HD

Search terms included:
Title: Huntington's disease, Chorea, Huntington, Huntington Chorea
Exp. Physical therapy modalities, physical therapy .mp., physiotherapy .mp., exp. Rehabilitation/ rehabilitation .mp., exp occupational therapy/ occupational therapy .mp.
Physical activity .mp., exp exercise therapy/ exercise therapy .mp., exp. Activities of daily living/ activities of daily living .mp., physical mobility .mp., exp muscle stretching exercises/ stretching .mp., exp respiratory therapy/ respiratory therapy .mp., flexibility .mp., exp range of motion/ range of movement .mp., exp upper extremity/ upper limb .mp., reaching .mp., reach .mp., grasping .mp., grasp .mp., reach to grasp .mp., exp posture/ posture .mp., balance .mp., exp accidental falls,/ falls .mp., exp equipment and supplies/ equipment .mp., seating .mp., positioning .mp., transfers .mp., exp functional status/ functional status .mp., exp gait/ gait .mp.

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Summary of Studies (up to 09/07/2008)

Abbreviations used in tables:

Abbreviation	Meaning
HD	Huntington's disease group
C	Control/ comparison group
M	Male
F	Female
↑	Increased/ improved
↓	Decreased/ decline

Binswanger C.
Physical therapy in Huntington disease.
Archives of Physical Medicine & Rehabilitation.
1980; 61(3):148.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Before-after observational Pilot study	Home-based	N=5 ? Confirmed diagnosis of HD	1 hour x 2 per week 4 weeks duration Delivered by a physiotherapist, individually tailored programmes to carry out self-supervised also. Components of intervention not specified but thought to include: • Neurophysiologic techniques • Range of motion exercises • Strengthening exercises • Gait rehabilitation • Breathing control		Not clearly defined States "standard physiotherapy tests of physical and functional capacity". Participant's alertness and ability/ willingness to participate in activities subjectively observed.	• Alertness • Ability/ willingness to engage in activities • Balance leading to safer ambulation

Lavers A.
An Account of a Weekly Activity Group with Huntington's Chorea Patients on a Long-stay Ward.
Occupational Therapy. 1981; 44:387-92.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Reflective report/ non experimental/ expert opinion	Long-stay inpatient psychiatric ward	N= 6 Confirmed diagnosis of HD	Multidisciplinary 1x per week 12-month duration. Physiotherapy intervention included: • Assisted walking • Mobility training • Mat exercises including prone lying, bridging, kneeling, crawling, long sitting, standing • Chair exercises including hand games, coordination and stretching activities • Weighted cuffs for upper limb coordination and assisted walking • Splinting • Positioning • Use of supportive devices		Not clearly defined/ none reported Physiotherapy specific aims: - Maintain mobility, balance and coordination - Prevent contractures - Increase social interaction - Reduce passive behaviour	No objective results • Week to week variability noted. • Despite the input, physical deliberation did occur • The group provided an aspect of maintenance therapy within advanced stage HD

Peacock IW.
A physical therapy program for Huntington's disease patients.
Clinical Management in Physical Therapy. 1987; 7(1):22.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Before-after observational study	Therapy centre for weekly sessions Home-based exercise for self-supervised intervention	N=10 Early to mid-stage HD Community dwelling	1 x per week 12 weeks duration + 3 booster sessions held monthly Total session duration: 2hrs Exercise duration: 45 mins Therapist led intervention: Relaxation followed by exercise Exercise individualised addressing maintenance of: • Functional ability • Flexibility • Coordination • Balance • Breathing control • Strength. Participants also given a list of exercises to carry out at home self-supervised/ family-supervised.		Set, validated outcome measures not included. 10 tests including: -Range of motion/ flexibility -Strength Co-ordinated and reciprocal movement -Standing, one foot and kneeling balance -Breathing volume and control	All participants improved on 7+/10 functional tests All participants: • ↑ Flexibility • ↑ Elements of coordination • ↑ Breathing control and volume 9 participants: • ↑ Standing balance • ↑ Strength

Sheaff F.
Hydrotherapy in Huntington's disease.
Nursing Times. 1990; 86(4):46-9.

Design	Setting	Participant		Intervention	Outcome Measures	Results
Single case study Non experimental	In patient ward	N=1 50 year old male Advanced stage HD • An emaciated frame • Difficulties feeding • Dysarthria • Incontinence • Difficulties walking • Chorea +		1x per week Duration not stated Physiotherapist supervised hydrotherapy sessions with gentle exercise	Not clearly defined • Effect on chorea • Participant's adherence • Participant's view on intervention • Ability to exercise in the water	• ↓ Chorea within water, and immediately following therapy • Participant able to gently exercise within the water • Good adherence – only one session missed due to illness. • Participant appeared to enjoy sessions

Quinn L, Rao A.
Physical therapy for patients with Huntington disease: current perspectives and case report.
Neurology Report. 2002; 26(3):145-53.

Design	Setting	Participant		Intervention	Outcome Measures	Results
Single case study Observational	Home-based	N=1 49 year old male Diagnosed with HD 17 years ago Previous referrals to physiotherapy for functional decline and falls (sometimes leading to injury) Participant on stable medication		5 x per week 14 weeks (only 12 weeks of therapy) 35 mins per session • Video based exercise designed to reduce impairment and promote balance, coordination and flexibility (unable to obtain content from online journal) • Physiotherapist instructed stretches, one leg balance, and tandem walking, and upper limb ball exercises. Encouraged use of walking stick and foot orthotics	• SF-36 • Number of falls • Modified falls scale • Berg Balance Scale • Self paced/ fast paced gait speed • UHDRS Motor section • Physical examinations of posture, range of movement, strength	↓ Level of disability (SF-36) ↓ Number of falls ↑ Walking speed Improvement in: • Falls efficacy scale • Berg balance scale (+9 points) • UHDRS Motor (dystonia, chorea and bradykinesia) Patient subjectively reported enjoying the intervention and a wish to continue.

Quinn L, Rao A. **Physical therapy for patients with Huntington disease: current perspectives and case report.** Neurology Report. 2002; 26(3):145-53.

Components of the video exercise program for individuals with HD (35 minutes total time)		
Exercise type		**Specific exercises**
Warm up *Each exercise performed 10 times*		deep breathing neck flexion, extension/ rotation arm circles forward and backward squatting reaching up to "sky", alternating arms reaching to each side, crossing arms across body reach to floor, keeping knees slightly bent
Leg exercises (standing) *Each exercise performed 10-15 times*		knee lifts both legs* kicks to front both legs* squats * walk forwards and back walk to side (side step) jump in place *
Arm exercises (standing) *Each exercise performed 10-15 times both arms*		using 2 lb. weights, or heavy cans overhead press large arm circles biceps curls theraband exercises horizontal abduction shoulder flexion/extension
Floor exercises – stretching and strengthening		hamstring stretch (hold 30 seconds x 3 each leg) inner thigh stretch (hold 30 seconds x 3) sit ups (20 times) straight leg raises (15 each leg) prone press ups (10 times) push ups (15 times) cat camel
Cool down		Repeat warm up
		* Participants are instructed to hold onto chair for support if needed

Zinzi P, Salmaso D, De Grandis R, Graziani G, Maceroni S, Bentivoglio A, et al. **Effects of an intensive rehabilitation programme on patients with Huntington's disease: a pilot study.** Clinical Rehabilitation. 2007; 21(7):603-13.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Before-after Pilot study	Inpatient rehabilitation centre	N=40 M=17 & F=23 • Confirmed diagnosis of HD • Early to mid stage • Absence of severe psychiatric disease • Acceptable cognitive ability • All symptomatic	6x per week 3 weeks x3 per year for 2 years Per week: 8hrs per day= 5 days 4hrs per day = 1 day Individualised treatment programmes Exercise performed both individually and in groups Multidisciplinary led Physiotherapy Intervention included: • Respiratory intervention to ↑ breathing efficiency and coughing • Facial exercise • Whole body exercises in lying standing and sitting for: • Gait • Balance • Transfer training • Strengthening • Flexibility • Coordination • Postural training Common gym equipment used i.e. treadmill, bike, wall bars, balance board, step, free weights		At baseline for each admission: • Zung depression scale • Mini mental status examination (MMSE) • Barthel Index (ADL) Baseline and post-admission: • Tinetti scale (balance) • Physical Performance Test (PPT)	No significant change in Zung, MMSE, Barthel Index at each admission over the two years – no cognitive or functional decline Motor performance showed significant improvement from baseline at end of admission for each admission Mean Tinetti score ↑: +4.7 Mean PPT score ↑: +5.21 No carry over effect of improvement to next admission Tinetti and PPT scores (Motor function) maintained at baseline level over the two years – no motor decline Note: only N= 11/ 40 completed all 6 admissions

Busse M, Khalil H, Quinn L, Rosser A.
Physical Therapy Intervention for People with Huntington Disease.
Phys. Ther. 2008; 88 (7) 820-831

Design	Setting	Participant	Intervention		Outcome Measures	Results
Qualitative design	UK wide	Physiotherapists working with HD people	Interviews (n=8); questionnaires (n=49)		N/A; thematic analysis of interview and questionnaire data	Main issues that emerged from the data were classified into three sub-themes: (1) There is insufficient use of routine physical therapy-related outcome measures at different stages of HD; (2) There is under-utilization of physical therapy services in managing people with HD (particularly in the early stages) and (3) Management of falls and mobility-deficit progression is a key treatment aim for people with HD.

Zinzi P, Salmaso D, Frontali M.
Patient's and caregivers' perspectives: assessing an intensive rehabilitation programme and outcome in Huntington's disease.
J Public Health. 2009; Published online;
DOI 10.1007/s10389-009-0252-y

Design	Setting	Participant	Intervention		Outcome Measures	Results
Postal survey		People with HD who had completed at least one course of inpatient rehabilitation protocol within a pilot study in the previous 3 years.	Questioners (N=37) The aim is to evaluate patient's and caregiver's perspectives of an intensive rehabilitation program.		N/A; questionnaire data	General improvement after discharge was perceived by all of the respondents. Improvements were reported on gait, balance, and fall reduction. Duration of benefit was estimated to last from 1 to 3 months by 71% of the respondents. Majority of the respondents reported the patient's intention to continue with the rehabilitation programme in the future. From the free comments of the caregivers, the following themes have emerged: • Better knowledge of HD • Better sense of control in patient management • Empowerment in relationship with the neurologist and family doctor. • Increase hope for the future of their children at risk.

Hausdorff JM, Cudkowicz ME, Firtion R, Wei JY, Goldberger AL.
Gait variability and basal ganglia disorders: stride-to stride variations of gait cycle timing in Parkinson's disease and Huntington's disease.
Mov Disord. 1998; 13(3):428-37.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	(N=20) HD (N=15) PD (N=20) Healthy controls	Quantitative measures of gait were assessed using force sensitive insoles.		Quantitative measures of gait.	All measures of gait variability were significantly increased in PD and HD compared to control. In subjects with PD, gait variability was two times that observed n controls. In subjects with PD, gait variability was three times that observed n controls. The degree of gait variability in both conditions was correlated with disease severity. Gait speed was lower in PD compared to controls. Gait speed in HD was similar to the observed gait speed in controls.

Thaut MH, Miltner R, Lange HW, Hurt CP, Hoemberg V.
Velocity modulation and rhythmic synchronization of gait in Huntington's disease.
Movement Disorders. 1999; 14(5):808-19.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Before-after Observational study	Laboratory based	N=27 (M=13 & F=14) Confirmed diagnosis of HD Mean age: 47 + 10.7 yrs Mean disease duration: 7.3+3.3yrs Shoulson-Fahn scores: Mean chorea: 1.37+0.61 Mean disability: 1.28+0.68 19 participants medicated	20m gait analysed Uncued (self-paced): 4 gait trials 1) Normal speed pre-test baseline 2) Slower than baseline 3) Faster than baseline 4) Normal speed 1st post-test Cued: Rhythmic Auditory Stimulation (RAS) 1) RAS metronome at 10% slower than baseline 2) RAS metronome at 10-20% faster than baseline 3) RAS music faster than baseline 4) Normal speed 2nd post-test		Gait velocity Cadence Stride length Swing symmetry Cadence asynchrony to RAS	Baseline: All parameters abnormal e.g. ↓ velocity, stride length, and cadence compared to aged matched healthy comparison Uncued: Slow N=25/27 able to ↓ velocity by 25.5%. Gait symmetry ↓ Fast N=19/27 able to ↑ velocity by 24.3%. Gait symmetry ↑ Cued: Slow N=21/27 able to ↓ velocity by 20%. Gait symmetry stable Fast N=23/27 able to ↑ velocity by 26%. Gait symmetry ↑ Music N=17/27 able to ↑ velocity slightly by 9.3% - difficulties modulating velocity Synchronisation Participants were able to synchronise to RAS, especially music Main finding: Participants able to modulate velocity with and without cue present. However, whilst gait remained unchanged between baseline and 1st post-test, velocity significantly improved between baseline and 2nd post-test – TRAINING EFFECT WITH CUE Ability to ↑ gait velocity with / without cue ↓ with disease severity

Churchyard AJ, Morris ME, Georgiou N, Chiu E, Cooper R, Iansek R.
Gait dysfunction in Huntington's disease: parkinsonism and a disorder of timing. Implications for movement rehabilitation.
Advances in Neurology. 2001; 87:375-85.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Before-after observational study	Laboratory based	HD group (HD) N=20 Confirmed diagnosed of HD Comparison group (C): N=16 Age matched healthy	5 x 10 m walking trials for each condition Self-paced walking: 1) Preferred 2) Slow 3) Fast Externally triggered: Auditory cue (metronome) Attentional distraction (dual tasking): Walking whilst counting backwards		• UHDRS • Cognitive, oral and mood measures (not discussed here) • Gait velocity (m/min) • Stride length (m) • Cadence (steps/ min) • Duration of double support phase	UHDRS motor score correlated with severity of hypokinesia Preferred gait (compared to comparison group): ↓ velocity ↓ stride length ↓ cadence Velocity, stride length, cadence ↑ variability ++ Slow and fast walking (internal cue) (compared to comparison group) HD group could modify velocity and stride length but still ↓ velocity ↓ stride length Velocity, stride length, cadence ↑ variability ++ External cue (Metronome) Difference between groups in velocity and cadence ↓ but not "normalised" Cadence modulated by metronome but not "normalised" Velocity, stride length, cadence ↑ variability ++ Distraction (Dual tasking) HD group further ↓ velocity and cadence++

Rao AK, Quinn L, Marder KS.
Reliability of spatiotemporal gait outcome measures in Huntington's disease.
Mov Disord. 2005; 20(8):1033-7.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	(N=12) confirmed diagnosis of HD (N=12) healthy matched controls	Aim: Evaluating sensitivity and test-restest reliability of gait outcome measures in HD using the GaitRite instrument. Each subject underwent two separate assessment sessions. Each subject performed two trials at each assessment session.		**Gait measures:** • **Gait velocity** • **Cycle time** • **Stride length** • **Cadence** • **Base of support**	Test-retest reliability of outcome measures Test retest variability of GaitRite instrument was very high for all gait measures ((ICC= 0.86 to 0.95) and (CV=.042 to 0.102)) Comparison of HD and control subjects GaitRite is sensitive enough to distinguish between control and HD subjects.

Bilney B; Morris ME, Churchyard A, Chiu E, Georgiou-Karistianis N.
Evidence for a disorder of locomotor timing in Huntington's disease.
Move Disord. 2005; 20(1):51-7.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	(N=30) confirmed diagnosis of HD (N=30) healthy matched controls	Gait parameters were measured using a computerized foot-switch system at two conditions 1- Uncued walking at self selected slow, preferred and fast speeds 2- Cued walking: walking in time to a metronome at 80 and 120 beats per minute		• Gait speed • Cadence • Stride length • Double limb support	preferred and fast walking (comparison to control group) ↓ velocity ↓ cadence All conditions (comparison to control group) ↓ stride length ↑ Variability of cadence, stride length, step time Cueing (walking to metronome) HD group have difficulty in synchronizing foot step time to the external auditory cue

Delval A, Krystkowiak P, Blatt JL, Labyt E, Bourriez JL, Dujardin K, et al.
Role of hypokinesia and bradykinesia in gait disturbances in Huntington's disease: a biomechanical study.
J Neurol 2006; 253:73-80.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	HD Group (HD): N=15 Confirmed diagnosis of HD (early stage) Comparison group (C): N=15 Age and gender matched No neurological deficits	Hypokinesia was studied in terms of both spatial (decrease in stride length) and angular gait parameters (decrease in joint ankle range), whereas hyperkinesia was characterized by an increase in joint ankle range. Bradykinesia (defined by a decrease in gait velocity) was also assessed in terms of temporal parameters (cadence, stride time)		Multiple outcome measures used including kinematics, spatiotemporal and joint angles	↓ velocity and cadence ↑ stride time (i. e. bradykinesia) for HD no clear ↓ in stride length angle analysis revealed the coexistence of hyperkinesia and hypokinesia in HD

Delval A, Krystkowiak P, Blatt JL, Labyt E, Bourriez JL, Dujardin K, et al.
A biomechanical study of gait initiation in Huntington's disease.
Gait & Posture. 2007; 25(2):279-88.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	HD Group (HD): N=15 Confirmed diagnosis of mild to moderate HD Mean age: 47+11.2 yrs Mean time since onset: 5.8+3.6 years Mean Total Functional Capacity (TFC): 10.9+1.6 Motor UHDRS: 28.7+22 Medication not altered during study Comparison group (C): N=15 Age and gender matched No neurological deficits	Gait initiation on a force platform + 4 to 5 following steps 1) Externally triggered Auditory cue (beep) to signify gait initiation once participant ready. 2) Self triggered Participant began when ready at fastest speed possible 3-5 trials for each condition		Multiple outcome measures used including kinematics, spatiotemporal and joint angles Selection of outcome measures: Centre of pressure (COP) trajectories First step speed (m/s) First step length (m) First step duration (s) Second step speed (m/s) Second step length (m) Second step duration (s) Anticipatory postural adjustments (APA) – duration of heel off for starting leg	1st and 2nd step speed and length ↓ and step duration ↑ for HD compared to C for both conditions External cue reduced between group differences improving 1st and 2nd step speed and duration for HD External cue minimised between group differences for COP trajectories, reducing deterioration in movement preparation HD APA= C group APA when external cue was present

Delval A, Krystkowiak P, Delliaux M, Dujardin K, Blatt J, Destée A et al.
Role of attentional resources on gait performance in Huntington's disease.
Movement Disorders. 2008; 23 (5) 684-689.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	HD Group (HD): N=15 People had to be able to walk unaided for at least 10 m without stopping or falling. People had also to be able to perform the cognitive tasks. Mean age: 43.9+ 9.8 Mean time since onset: 4.9 +3.2 years Mean Total Functional Capacity (TFC): 9+ 2.5 Motor UHDRS: 42+ 17.1 Medication not altered during study Comparison group (C): N=15 Age and gender matched No neurological deficits	Kinematic spatial, temporal, and angular gait measurements were automatically recorded by means of a video motion system. Subjects walked a distance of 10 m 1) Self-Selected Walking Speed or Free Gait 2) Motor Dual Task (Tray With Four Glasses). 3) Cognitive Dual Task: Backward Count 10 gait cycles were taken into account for each subject (five right leg cycles and five left leg cycles) for each condition.		Stride length (m), velocity (m/s)] and temporal kinematic Gait parameters [cadence (steps/ min), velocity (m/s), ratio of single limb support time to double limb support time]. Kinematic angle data [sagittal plane excursions hip, knee, and ankle angles]	Dual-task performance resulted in a greater reduction in walking speed, stride length, and cadence in people with HD than in healthy control subjects. A cognitive dual task resulted in a worsening of all spatiotemporal kinematic parameters (in contrast with the motor dual task), suggesting that a cognitive-motor dual task may generate more interference than a dual-motor task.

Delval A; Krystkowiak P; Delliaux M; Blatt JL; Derambure P; Destée A; Defebvre L.
Effect of external cueing on gait in Huntington's disease.
Movement Disorders. Early View

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	Fifteen HD people; age 43.9 ± 9.8; matched healthy controls	Free Gait (FG), Metronome 120% (M120) Gait + Motor Task (Carrying a Tray with Four Glasses), Gait + Motor Task, Metronome at 100%, Gait + Motor Task, Metronome at 120% Gait + Cognitive Task: Counting Backwards, Gait + Cognitive Task with Metronome at 100%, Gait + Cognitive Task with Metronome at 120%		Kinematic spatial, temporal, and angular gait measurements (VICON system)	1) the effect of a metronome on free gait: lower cadence, stride length, and gait speed in HD people than in controls 2) effect of a metronome on gait plus a motor task and cognitive task: higher gait speed and cadence (but not greater stride length) in controls compared to HD people 3) No statistically significant positive impact of a metronome on gait parameters in HD people performing dual tasks, although trend towards an improvement

Grimbergen Y; Knol M; Bloem B; Kremer B; Roos R; Munneke M.
Falls and gait disturbances in Huntington's disease.
Movement Disorders. 2008; 23(7): 970-976

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	45 early to mid-stage HD people; 27 healthy age-matched controls.			Falls; UHDRS scores; Quantitative measures of balance (using angular velocity sensors) Gait (using a pressure-sensitive walkway: GaitRite)	Twenty-seven people (60%) reported two or more falls in the previous year; Fallers showed significantly higher scores for chorea, bradykinesia and aggression, as well as lower cognitive scores. HD people had a decreased gait velocity and decreased stride length compared to controls and in fallers these were all significantly greater compared to HD non-fallers.

Rao A; Muratori L; Louis E; Moskowitz C; Marder K.
Spectrum of gait impairments in presymptomatic and symptomatic Huntington's Disease.
Movement Disorders. 2008; 23 (8): 1100-1107

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	Presymptomatic mutation carriers (PMC) (n =15), symptomatic HD subjects (SHD) (n = 30) and healthy controls (n = 20)	Preferred walking speed		Gait (using a pressure-sensitive walkway: GaitRite); UHDRS	Decrease in gait velocity with increasing disease severity; gait impairments were correlated with predicted years to onset in PMC and demonstrated high sensitivity and specificity in distinguishing between controls and mutation carriers.

Curra A, Agostino R, Galizia P, Fittipaldi F, Manfredi M, Berardelli A.
Sub-movement cueing and motor sequence execution in people with Huntington's disease.
Clinical Neurophysiology. 2000 Jul; 111(7):1184-90.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	HD group: N=7 (M=4& F=3) Mean age: 55.9+9.1yrs Mean disease duration: 4.5+2.1yrs Comparison group (C): N=7 (M=5&F=2) Mean age: 55.5+9.1yrs	Free "zigzag" arm movement joining targets on screen Blocks of 10 trials each, random order Self initiated sequence Move when ready as fast as possible Externally triggered sequence– visual cue: Targets change colour consecutively – move when change colour		Sub movement time (SMT)- movement time between targets Total Movement Time (TMT) – time to complete sequence Movement amplitude (MA) Externally triggered only: Reaction time (RT) Self initiated only: Duration of pauses	SMT: HD slower than C regardless of condition Both HD and C slower during externally triggered condition than self initiated condition TMT: HD slower than C regardless of condition Both HD and C slower during externally triggered condition than self initiated condition Note: SMT and MT differed less between cued/ self initiated conditions for HD than C. This indicates that the HD group had more difficulties with self-initiated, internally cued movement? RT: HD showed longer RT than C Duration of pauses: No significant differences between groups or conditions – HD group still able to sequence movement

Busse ME, Hughes G, Wiles CM, Rosser AE.
Use of hand-held dynamometry in the evaluation of lower limb muscle strength in people with Huntington's disease.
J Neurol. 2008;255(10):1534-40.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational design	Hospital clinic	(N=20) confirmed diagnosis with HD (N=20) healthy control subjects	Isometric muscle strength of 6 lower limb muscle groups was measured using a hand held dynamometer. Reliability and validity of muscle testing were calculated.		Muscle strength Within session reliability of muscle strength testing using intra-class correlation coefficient Construct validity of muscle testing.	Reliability of strength testing Reliability of strength testing was excellent (ICC 0.86 to 0.98). Validity of strength testing • People with HD had on average about half the strength of healthy matched controls. • UHDRS motor scores and strength scores were significantly correlated providing indication of further validity of strength testing in HD.

Busse ME, Wiles CM, Rosser A.
Mobility and falls in people with Huntington's disease.
J.Neurol. Neurosurgery. Psychiatry. 2009; 80:88-90.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational design	Hospital clinic	(N=20) confirmed diagnosis with HD	Aims: 1. Estimating the frequency of falls in HD. 2. Preliminary assessment of tools appropriate for assessing risk of falls in HD.		• Balance (BBS &TUG) • Walking speed (10 m walking test) • Fall history (data about falls and stumbles in the previous 12 months).	Differences between ''recurrent fallers'' (≥2 falls/year) and ''non-fallers'' (≤ 1 fall/year) for the range of outcome measures was calculated. Fall history • 41.6% of the patients reported ≤ 1 fall in the previous 12 months (non-fallers). • 58.3% of patients reported ≥2 falls in the previous 12 months (recurrent fallers). Walking speed • Recurrent fallers walked less and slower than the non-fallers. Balance • Recurrent fallers have worse balance scores (as were measured by BBS) than the non fallers. • Recurrent fallers have higher TUG scores than the non fallers. Risk of falls • TUG &BBS has been identified to be significant predictors of falls in this population. • People with HD have increased risk of falls if TUG scores were ≥14 or BBS scores≤ 40.

Rao AK, Muratori L, Louis ED, Moskowitz CB, Marder KS.
Clinical measurements of mobility and balance impairments in Huntington's disease: Validity and responsiveness.
Gait and posture. 2009; 29: 433-436.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational study	Laboratory based	(N=30)) confirmed diagnosis with HD	Aim: To examine validity and responsiveness of three clinical tests of mobility and balance in HD. The tests include: • Berg balance scale (BBS) • Timed Up and Go (TUG) test • Functional reach (FRT) test		• Balance and mobility were assessed using: • Berg balance scale (BBS) • Timed Up and Go (TUG) test • Functional reach (FRT) test • Functional limitations were assessed by using : • UHDRS • Total Functional Capacity (TFC) scale • Huntington's disease-activities of daily living scale (HD-ADL) • Symptom onset (years when the subject first observed the symptoms were recorded) • Quantitative gait data; (collected using pressure-sensitive walkway: GaitRite)	FRT and BBS scores were correlated with five quantitative gait measures. TUG scores were correlated with eight quantitative gait measures. All tests were correlated with indications of functional limitations. All tests were responsive to disease severity.

Tian J, Herdman SJ, Zee DS, Folstein SE.
Postural stability in patients with Huntington's disease.
Neurology. 1992;42(6):1232-8.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational design	Laboratory based	1-N=20 HD patients 2- N=20 matched healthy controls	• Moving platform posturography was used • The six conditions were described as follows: 1 all cues available 2 No vision 3 Vision attenuated 4 Somatosensory attenuated 5 Somatosensory attenuated+no vision 6 Both vision and somatosensory are attenuated		1 AP sway Difference between maximum anterior and maximum posterior displacement of the COM in each trial. 2 Motor strategy used to maintain balance Relative amount of ankle sway and hip movement to maintain balance was calculated. 3 Responses to imposed perturbations 1 Latency: time between onset of translation and onset of motor response. 2 strength: rate of change of projection of vertical forces	HD increased AP sway when standing on stationary platform compared to control. The sway in HD is increased when the vision is eliminated compared to control The sway in HD is increased when the proprioceptive input is attenuated compared to control. With external pertrusion: The corrective postural response in HD has significantly delayed compared to control.

Louis ED, Lee P, Quinn L, Marder K.
Dystonia in Huntington's disease: prevalence and clinical characteristics.
Mov Disord. 1999;14(1):95-101.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational design	Laboratory based	N=42 Confirmed diagnosed of HD	Patients underwent a standardized 5.5 minutes of video examination. • 45 sec; patient is asked to sit and read a passage aloud • 45sec;patient is asked to sit and face the camera • 4 minutes; patient is asked to walk. Two neurologists reviewed the tape and rated the dystonia using a scale developed for the study.		• Prevalence of dystonia • Clinical features of dystonia 1. Types 2. Severity 3. Constancy	Prevalence of dystonia of any severity was about 95% 57% of patients has dystonia at least at one part of the body 16% had severe and constant dystonia The mean severity was between 1 (mild) and 2 (moderate) The mean constancy was between 2 (present less than half of the time) and 3 (present more than half of the time) The most prevalent types are: • Internal rotation of the shoulder. • Fist clinching • Increased knee flexion • Increased foot inversion

Quinn L, Reilmann R, Marder K, Gordon AM.
Altered movement trajectories and force control during object transport in Huntington's disease.
Movement Disorders. 2001 May; 16(3):469-80.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Observational Study	Laboratory Based	HD group: N=12 Confirmed diagnosis of HD Mean age 55yrs -Minimal upper limb rigidity -High cognitive ability Comparison group (C): N=12 Age matched	In a seated position with a table in front, participants: 1) Reached forward 10cm 2) Grasped and lifted the experimental object with a precision grip 3) Transported it 25cm forward 4) Placed it down on a marker on the table. Task performed at preferred speed Experimental object weight modified: 200g, 400g, 800g 5 practice trials for each weight 5 test trials for each weight		-Contact time (thumb and finger contact with object) Preload phase (transition from reach-to-grasp to grasp-to-lift) Loading phase (increase of grip and load force) Transport time (movement from start to finish position) Peak transport velocity Unloading phase (replacement of object and release of one finger) Release phase (release of two fingers) Transport grip force Hand path	<u>Contact time:</u> ↑ in HD compared to C ↓ with object weight for HD <u>Preload phase:</u> ↑ in HD compared to C regardless of object weight <u>Loading phase:</u> No significant difference between groups or object weight conditions <u>Transport time:</u> ↑ in HD compared to C ↓ in HD group with ↑ object weight <u>Peak velocity:</u> Similar for both HD and C group <u>Grip force:</u> ↑ for HD compared to C group regardless of object weight conditions <u>Grip force variability:</u> Significantly ↑ in HD group within and between trials. Variability ↓ at maximum object weight 800g <u>Hand Path:</u> Straight in C group, curvilinear in HD group. This did not effect the accuracy of final object position For HD group ↑ object weight improved linearity of path <u>Unload and release phase:</u> HD group comparable to C group.

Leng TR, Woodward MJ, Stokes MJ, Swan AV, Wareing L, Baker R.
Effects of multisensory stimulation in people with Huntington's disease: a randomized controlled pilot study.
Clinical Rehabilitation. 2003; 17(1):30-41.

Design	Setting	Participant	Intervention		Outcome Measures	Results
Randomised Controlled Trial Pilot study	Inpatient residential unit	Intervention group (MSE) N=6 Control group (C) N=6 All with a confirmed diagnosis of HD 5yrs+ All advanced (stage V) disease requiring total care	2x per week 4 weeks 30min per session Participants treated individually Multi-sensory stimulation (MSE): Intervention Equipment used: 1) Visual: projector, fibre optic sprays, bubble tubes, kaleidoscope 2) Tactile: vibrating cushion, balls, fabrics, handheld massager, water balloon 3) Auditory: relaxation music 4) Olfactory: Aromatherapy Relaxation: Control: Listened to music Therapist read		12 weeks: 4 week baseline, intervention, and follow up Sustained effects: 2wk intervals • Rehabilitation evaluation scale (REHAB) • Behaviour and mood disturbance scale (BMD) Immediate effects: Before, during and immediately after sessions • Interact (behaviour assessment) Measured 3 days per wk: • Heart Rate (HR) • Blood pressure (BP) • Respiratory rate (RR) • St Hans rating scale (involuntary movement) (SHRS)	N=2 withdrawn due to medical complications Sustained effects (therapeutic effect) No significant changes for REHAB, BMD, SHRS, BP, HR and RR during baseline, intervention or follow up for either group. Immediate effect (leisure) Interact: MSE group showed significant improved in mood and stimulation compared to control group. This effect was cumulative over sessions No change from before, to during and after sessions for either group in SHRS, BP, HR and RR Summary: MSE= useful leisure activity, providing management resource for complex neuro-disability

EXPERT OPINION: Study Summary

Imbriglio S, Peacock IW.
Huntington's disease at mid-stage.
Clinical Management. 1992; 12 (5):62-72.

- Physiotherapy is beneficial in Huntington's disease (HD), improving/ stabilising motor ability, preventing contractures, and adapting the environment to maintain independence and safety.
- A physiotherapy evaluation should include: range of motion, muscle strength, leg length, trunk mobility, standing and sitting balance, posture, sensation, cardiopulmonary status, pain, transfers, gait, the need for special equipment, the need for special tests.
- Exercise and rehabilitation should start early in the disease, before many people feel they need it.
- As People with HD will be aware of the degenerative nature of their condition and may doubt the need for rehabilitation, support from the physiotherapist is required: explanation/ persuasion of the benefits of physiotherapy, provision of drop-in sessions, and support with remembering to attend sessions.
- The structure of the intervention is patient dependant – some will prefer group work to individualised sessions.
- Small group sessions provide therapy along with support and social interaction, yet still allow the patient to be individually managed
- Interventions should focus on: coordination skills, ambulation skills, relaxation skills, and sensory awareness.
- Therapy sessions can be recreational as well as therapeutic involving balls games etc.
- Focus on what participants can do and maintaining current levels of function.
- Decisions should be made collaboratively between the therapist and participant.
- A key role of the physiotherapist in HD is the assessment of safety during physical functioning.
- Another key role is communication, support and collaboration with the family members.

Appendix Two: Outcome Measures

WHOQOL-BREF

The World Health Organization Quality of Life (WHOQOL)-BREF

WHOQOL-BREF

The following questions ask how you feel about your quality of life, health, or other areas of your life. I will read out each question to you, along with the response options. **Please choose the answer that appears most appropriate.** If you are unsure about which response to give to a question, the first response you think of is often the best one.

Please keep in mind your standards, hopes, pleasures and concerns. We ask that you think about your life **in the last four weeks.**

		Very poor	Poor	Neither poor nor good	Good	Very good
1.	How would you rate your quality of life?	1	2	3	4	5

		Very dissatisfied	Dissatisfied	Neither satisfied nor dissatisfied	Satisfied	Very satisfied
2.	How satisfied are you with your health?	1	2	3	4	5

The following questions ask about **how much** you have experienced certain things in the last four weeks.

		Not at all	A little	A moderate amount	Very much	An extreme amount
3.	To what extent do you feel that physical pain prevents you from doing what you need to do?	5	4	3	2	1
4.	How much do you need any medical treatment to function in your daily life?	5	4	3	2	1
5.	How much do you enjoy life?	1	2	3	4	5
6.	To what extent do you feel your life to be meaningful?	1	2	3	4	5

		Not at all	A little	A moderate amount	Very much	Extremely
7.	How well are you able to concentrate?	1	2	3	4	5
8.	How safe do you feel in your daily life?	1	2	3	4	5
9.	How healthy is your physical environment?	1	2	3	4	5

The following questions ask about how completely you experience or were able to do certain things in the last four weeks.

		Not at all	A little	Moderately	Mostly	Completely
10.	Do you have enough energy for everyday life?	1	2	3	4	5
11.	Are you able to accept your bodily appearance?	1	2	3	4	5
12.	Have you enough money to meet your needs?	1	2	3	4	5
13.	How available to you is the information that you need in your day-to-day life?	1	2	3	4	5
14.	To what extent do you have the opportunity for leisure activities?	1	2	3	4	5

		Very poor	Poor	Neither poor nor good	Good	Very good
15.	How well are you able to get around?	1	2	3	4	5

		Very dissatisfied	Dissatisfied	Neither satisfied nor dissatisfied	Satisfied	Very satisfied
16.	How satisfied are you with your sleep?	1	2	3	4	5
17.	How satisfied are you with your ability to perform your daily living activities?	1	2	3	4	5
18.	How satisfied are you with your capacity for work?	1	2	3	4	5
19.	How satisfied are you with yourself?	1	2	3	4	5

20.	How satisfied are you with your personal relationships?	1	2	3	4	5
21.	How satisfied are you with your sex life?	1	2	3	4	5
22.	How satisfied are you with the support you get from your friends?	1	2	3	4	5
23.	How satisfied are you with the conditions of your living place?	1	2	3	4	5
24.	How satisfied are you with your access to health services?	1	2	3	4	5
25.	How satisfied are you with your transport?	1	2	3	4	5

The following question refers to how often you have felt or experienced certain things in the last four weeks.

		Never	Seldom	Quite often	Very often	Always
26.	How often do you have negative feelings such as blue mood, despair, anxiety, depression?	5	4	3	2	1

Do you have any comments about the assessment?

_____

_____

[The following table should be completed after the interview is finished]

		Equations for computing domain scores	Raw score	Transformed scores*	
				4-20	0-100
27.	**Domain 1**	(6-Q3) + (6-Q4) + Q10 + Q15 + Q16 + Q17 + Q18 □ + □ + □ + □ + □ + □ + □	a. =	b:	c:
28.	**Domain 2**	Q5 + Q6 + Q7 + Q11 + Q19 + (6-Q26) □ + □ + □ + □ + □ + □	a. =	b:	c:
29.	**Domain 3**	Q20 + Q21 + Q22 □ + □ + □	a. =	b:	c:
30.	**Domain 4**	Q8 + Q9 + Q12 + Q13 + Q14 + Q23 + Q24 + Q25 □ + □ + □ + □ + □ + □ + □ + □	a. =	b:	c:

* See Procedures Manual, pages 13-15

6-Minute Walk Test

Description: The 6-Minute Walk test is a measure of endurance.
Equipment: stopwatch, rolling tape measure, track/loop walkway
Instructions: Monitor vital signs before and after each test if indicated. Assure patient safety throughout the test. Give the same verbal instructions each time. "When I say 'go', I want you to walk around this [track]. Keep walking until I say 'stop' or until you are too tired to go any further. If you need to rest, you can stop until you feel ready to go again. I am interested in measuring how far you can walk. You can begin when I say 'go'." Time the subject for 6 minutes, then say 'stop'. Measure the distance walked. Also document distance walked at 1 and 3 minutes.

Stop testing based on the following criteria:
1. c/o angina symptoms (chest pain or tightness)

2. Any of the following symptoms:
a. Light-headedness
b. Confusion
c. Ataxia, staggering unsteadiness
d. Pallor
e. Cyanosis
f. Nausea
g. Marked dyspnea
h. Unusual fatigue
i. Signs of peripheral circulatory insufficiency
j. Claudication or other significant pain
k. Facial expressions signifying distress

3. Abnormal cardiac responses
a. Systolic blood pressure drops > 10 mmHg
b. Systolic blood pressure rises < 250 mmHg
c. Diastolic blood pressure rises to > 120 mmHg
d. Heart rate drops more than 15 beats per minute (given the subject was walking the last minutes of the test versus resting)

Notify physician if test is terminated for any of the above reasons

10 metre Walk Test

A 14m walking area will be marked, with 2 m on either side allowed for starting and stopping of walking (acceleration and deceleration). Subjects will be asked to walk the length of the area at a comfortable walking speed. Subjects will be excluded from participating in this test if they are unable to ambulate without physical assistance. Assistive devices can be used. The total time to complete 10m, as well as the number of steps taken during those 10m will be recorded. Average gait speed (distance/time) and cadence (step/min) will then be calculated.

Timed "Up and Go"

Directions:
The timed "Up and Go" test measures, in seconds, the time taken by an individual to stand up from a standard arm chair (approximate seat height of 46 cm, arm height 65 cm), walk a distance of 3 meters (approximately 10 feet), turn, walk back to the chair, and sit down. The subject wears their regular footwear and uses their customary walking aid (none, cane, walker). No physical assistance is given. They start with their back against the chair, their arms resting on the armrests, and their walking aid at hand. They are instructed that, on the word "go" they are to get up and walk at a comfortable and safe pace to a line on the floor 3 meters away, turn, return to the chair and sit down again. The subject walks through the test once before being timed in order to become familiar with the test. Either a stopwatch or a wristwatch with a second hand can be used to time the trial.
Instructions to the patient:
"When I say 'go' I want you to stand up and walk to the line, turn and then walk back to the chair and sit down again. Walk at your normal pace."
Variations:
You may have the patient walk at a fast pace to see how quickly they can ambulate. Also you could have them turn to the left and to the right to test any differences.

Physical Performance Test

Testing Protocol:
Administer the test as outlined below. Subjects are given up to two chances to complete each item. Assistive devices are permitted for tasks 6 – 8.

1. Ask the subject, when given the command to "go" to write the sentence "whales live in the blue ocean." Time from the word "go" until the pen is lifted from the page at the end of the sentence. All words must be included and legible. Period need not be included for task to be considered completed.

2. Five kidney beans are placed in a bowl, 5 inches from the edge of the desk in front of the patient. An empty coffee can is placed on the table at the patient's non-dominant side. A teaspoon is place in the patient's dominant hand. Ask the subject on the command "go" to pick up the beans, one at a time and place each in the coffee can. Time from the command "go" until the last bean is heard hitting the bottom of the can.

3. Place a Physician's Desk Reference or other heavy book on a table in front of the patient. Ask the patient, when given the command "go" to place the book on a shelf above shoulder level. Time from the command "go" to the time the book is resting on the shelf.

4. If the subject has a jacket cardigan sweater, ask them to remove it. If not, give the subject a lab coat. Ask the subject, on the command "go" to put the coat on completely such that it is straight on their shoulders and then remove the garment completely. Time from the command "go" until the garment has been complexly removed.

5. Place a penny approximately1 foot from the patient's foot on the dominant side. Ask the patient, on the command "go" to pick up the penny from the floor and stand up. Time from the command "go" until the subject is standing erect with a penny in hand.

6. With subject in a corridor or in and open room, ask the subject to turn 360 degrees. Evaluate using the scale on PPT scoring sheet.

7. Bring subject to start on a 50 –foot walk test course (25 feet out and 25 feet back) and ask the subject, on the command "go" to walk to the 25-foot mark and back. Time from the command "go" until the starting line is crossed on the way back.

8. Bring subject to foot of stairs (nine to 12 steps) and ask subject, on the command "go" to begin climbing stairs until they feel tired and wishes to stop. Before beginning this task, alert the subject to the possibility of developing chest pain or shortness of breath and inform the subject to tell you if any of these symptoms occur. Escort the subject up the stairs. Time from the command "go" until the subjects' first foot reaches the top of the first flight of stairs. Record the number of flights (maximum is four) completed (up and down is one flight).

Scoring Sheet

	Task		Time	Scoring	Score
1.	**Write a sentence.** **(Whales live in the blue ocean.)**	Seconds		≤ 10 sec = 4 10.5-15 sec = 3 15.5 – 20 sec = 2 >20 sec = 1 unable = 0	
2.	**Simulated eating**	Seconds		≤ 10 sec = 4 10.5-15 sec = 3 15.5 – 20 sec = 2 >20 sec = 1 unable = 0	
3.	**Lift a book and put it on a shelf** **• Book PDR 1988: 5.5 lbs** **• Bed height 59 cm** **• Shelf height 118 cm** **• All sitting with feet on floor**	Seconds		≤ 2 sec = 4 2.5- 4 sec = 3 4.5 – 6 sec = 2 > 6 sec = 1 unable = 0	
4.	**Put on and remove a jacket** **Standing** **• Use of bathrobe;** **• button down shirt;** **• hospital gown.**	Seconds		≤ 10 sec = 4 10.5-15 sec = 3 15.5 – 20 sec = 2 >20 sec = 1 unable = 0	
5.	**Pick up a penny from floor.**	Seconds		≤ 2 sec = 4 2.5- 4 sec = 3 4.5 – 6 sec = 2 > 6 sec = 1 unable = 0	

	Task		Time	Scoring	Score
6.	**Turn 360 degrees**		Discontinuous steps = 0 Continuous steps = 2	Unsteady (grabs, staggers) = 0 Steady = 2	
7.	**50-foot walk test.** **Starting sitting for instructions.**	Seconds		≤ 15 sec = 4 15.5 – 20 sec = 3 20.5 – 25 sec = 2 >25 sec = 1 unable = 0	
8.	**Climb one flight of stairs (+)**	Seconds		≤ 5 sec = 4 5.5 – 10 sec = 3 10.5 – 15 sec = 2 >15 sec = 1 unable = 0	
9.	**Climb stairs (+)**		Number of flights of stairs up and down (maximum 4)		
	TOTAL SCORE (maximum 36 for nine-item, 28 for seven-item)				
	(*Round time measurements to nearest 0.5 seconds.) **(+ omit for 7 item test)**			9-item score	

Rivermead Mobility Index

1. Do you turn over from your back to your side without help?
2. From lying in bed, are you able to get up to sit on the edge of the bed on your own?
3. Could you sit on the edge of the bed without holding on for 10 seconds?
4. Can you (using hands and an aid if necessary) stand up from a chair in less than 15 seconds, and stand there for 15 seconds
5. Observe patient standing for 10 seconds without any aid
6. Are you able to move from bed to chair and back without any help?
7. Can you walk 10 metres with an aid if necessary but with no standby help?
8. Can you manage a flight of steps alone, without help?
9. Do you walk around outside alone, on pavements?
10. Can you walk 10 metres inside with no calliper, splint or aid and no standby help?
11. If you drop something on the floor, can you manage to walk 5 metres to pick it up and walk back?
12. Can you walk over uneven ground (grass, gravel, dirt, snow or ice) without help?
13. Can you get in and out of a shower/ bath unsupervised, and wash yourself?
14. Are you able to climb up and down four steps with no rail but using an aid if necessary?

15. Could you run 10 metres in 4 seconds without limping?
    (A fast walk is acceptable.)

TOTAL
Score 0 = No 1 = Yes

Barthel Index

Activity Score

FEEDING
0 = unable
5 = needs help cutting, spreading butter, etc., or requires modified diet
10 = independent

BATHING
0 = dependent
5 = independent (or in shower)

GROOMING
0 = needs to help with personal care
5 = independent face/hair/teeth/shaving (implements provided)

DRESSING
0 = dependent
5 = needs help but can do about half unaided
10 = independent (including buttons, zips, laces, etc.)

BOWELS
0 = incontinent (or needs to be given enemas)
5 = occasional accident
10 = continent

BLADDER
0 = incontinent, or catheterized and unable to manage alone
5 = occasional accident
10 = continent

TOILET USE
0 = dependent
5 = needs some help, but can do something alone
10 = independent (on and off, dressing, wiping)

TRANSFERS (BED TO CHAIR AND BACK)
0 = unable, no sitting balance
5 = major help (one or two people, physical), can sit
10 = minor help (verbal or physical)
15 = independent

MOBILITY (ON LEVEL SURFACES)
0 = immobile or < 50 yards
5 = wheelchair independent, including corners, > 50 yards
10 = walks with help of one person (verbal or physical) > 50 yards
15 = independent (but may use any aid; for example, stick) > 50 yards

STAIRS
0 = unable
5 = needs help (verbal, physical, carrying aid)
10 = independent

TOTAL (0–100):

The Barthel ADL Index: Guidelines

1. The index should be used as a record of what a patient does, not as a record of what a patient could do.
2. The main aim is to establish degree of independence from any help, physical or verbal, however minor and for whatever reason.
3. The need for supervision renders the patient not independent.
4. A patient's performance should be established using the best available evidence. Asking the patient, friends/relatives and nurses are the usual sources, but direct observation and common sense are also important. However direct testing is not needed.
5. Usually the patient's performance over the preceding 24-48 hours is important, but occasionally longer periods will be relevant.
6. Middle categories imply that the patient supplies over 50 per cent of the effort.
7. Use of aids to be independent is allowed.

References

Mahoney FI, Barthel D. "Functional evaluation: the Barthel Index." Maryland State Medical Journal 1965;14:56-61. Used with permission.
Loewen SC, Anderson BA. "Predictors of stroke outcome using objective measurement scales." Stroke. 1990; 21:78-81.
Gresham GE, Phillips TF, Labi ML. "ADL status in stroke: relative merits of three standard indexes." Arch Phys Med Rehabil. 1980; 61:355-358.
Collin C, Wade DT, Davies S, Horne V. "The Barthel ADL Index: a reliability study." Int Disability Study.1988; 10:61-63.

The Activities of Balance Confidence Scale (ABC)

Instructions to Participants:
For each of the following, please indicate your level of confidence in doing the activity without losing your balance or becoming unsteady from choosing one of the percentage points on the scale form 0% to 100%. If you do not currently do the activity in question, try and imagine how confident you would be if you had to do the activity. If you normally use a walking aid to do the activity or hold onto someone, rate your confidence as it you were using these supports. If you have any questions about answering any of these items, please ask the administrator.

For each of the following activities, please indicate your level of self confidence by choosing a corresponding number from the following rating scale:

0% 10 20 30 40 50 60 70 80 90 100%
no confidence completely confident

"How confident are you that you will not lose your balance or become unsteady when you…
1. …walk around the house? ____%

2. …walk up or down stairs? ____%

3. …bend over and pick up a slipper from the front of a closet floor ____%

4. …reach for a small can off a shelf at eye level? ____%

5. …stand on your tiptoes and reach for something above your head? ____%

6. …stand on a chair and reach for something? ____%

7. …sweep the floor? ____%

8. …walk outside the house to a car parked in the driveway? ____%

9. …get into or out of a car? ____%

10. …walk across a parking lot to the mall? ____%

11. …walk up or down a ramp? ____%

12. …walk in a crowded mall where people rapidly walk past you? ____%

13. …are bumped into by people as you walk through the mall?____%

14. … step onto or off an escalator while you are holding onto a railing? ____%

15. … step onto or off an escalator while holding onto parcels such that you cannot hold onto the railing? ____%

16. …walk outside on icy sidewalks? ____%

**Powell, LE & Myers AM. The Activities-specific Balance Confidence (ABC) Scale. J Gerontol Med Sci 1995; 50(1): M28-34*

Berg Balance Scale

Description:
14-item scale designed to measure balance of the older adult in a clinical setting.
Equipment needed:
- Ruler
- 2 standard chairs (one with arm rests, one without)
- Footstool or step
- Stopwatch or wristwatch
- 15 ft walkway

Completion:
Time: 15-20 minutes
Scoring:
A five-point ordinal scale, ranging from 0-4. "0" indicates the lowest level of function and "4" the highest level of function. TOTAL SCORE (0- 56)
Interpretation:
41-56 = low fall risk; 21-40 = medium fall risk; 0 –20 = high fall risk; < 36 fall risk close to 100%

ITEM DESCRIPTION SCORE (0-4)

1. Sitting to standing
2. Standing unsupported
3. Sitting unsupported
4. Standing to sitting
5. Transfers
6. Standing with eyes closed
7. Standing with feet together
8. Reaching forward with outstretched arm
9. Retrieving object from floor
10. Turning to look behind
11. Turning 360 degrees
12. Placing alternate foot on stool
13. Standing with one foot in front
14. Standing on one foot

Total _____

GENERAL INSTRUCTIONS

Please document each task and/or give instructions as written. When scoring, please record the lowest response category that applies for each item.

In most items, the subject is asked to maintain a given position for a specific time. Progressively more points are deducted if the time or distance requirements are note met, if the subject's performance warrants supervision, or if the subject touches an external support or receives assistance from the examiner. Subject should understand that they must maintain their balance while attempting the tasks. The choices of which leg to stand on or how far to reach are left to the subject. Poor judgment will adversely influence the performance and the scoring.

Equipment required for testing is a stopwatch or watch with a second hand, and a ruler or other indicator of 2, 5, and 10 inches. Chairs used during testing should be a reasonable height. Either a step or a stool of average step height may be used for item # 12.

Berg Balance Scale Scoring

1. SITTING TO STANDING

INSTRUCTIONS:
Please stand up. Try not to use your hand for support.
( ) 4 able to stand without using hands and stabilize independently
( ) 3 able to stand independently using hands
( ) 2 able to stand using hands after several tries
( ) 1 needs minimal aid to stand or stabilize
( ) 0 needs moderate or maximal assist to stand

2. STANDING UNSUPPORTED

INSTRUCTIONS:
Please stand for two minutes without holding on.
( ) 4 able to stand safely for 2 minutes
( ) 3 able to stand 2 minutes with supervision
( ) 2 able to stand 30 seconds unsupported
( ) 1 needs several tries to stand 30 seconds unsupported
( ) 0 unable to stand 30 seconds unsupported

If a subject is able to stand 2 minutes unsupported, score full points for sitting unsupported. Proceed to item #4.

3. SITTING WITH BACK UNSUPPORTED BUT FEET SUPPORTED ON FLOOR OR ON A STOOL

INSTRUCTIONS:
Please sit with arms folded for 2 minutes.
( ) 4 able to sit safely and securely for 2 minutes
( ) 3 able to sit 2 minutes under supervision
( ) 2 able to able to sit 30 seconds
( ) 1 able to sit 10 seconds
( ) 0 unable to sit without support 10 seconds

4. STANDING TO SITTING

INSTRUCTIONS:
Please sit down.
( ) 4 sits safely with minimal use of hands
( ) 3 controls descent by using hands
( ) 2 uses back of legs against chair to control descent
( ) 1 sits independently but has uncontrolled descent
( ) 0 needs assist to sit

5. TRANSFERS

INSTRUCTIONS:
Arrange chair(s) for pivot transfer. Ask subject to transfer one way toward a seat with armrests and one way toward a seat without armrests. You may use two chairs (one with and one without armrests) or a bed and a chair.
( ) 4 able to transfer safely with minor use of hands
( ) 3 able to transfer safely definite need of hands
( ) 2 able to transfer with verbal cuing and/or supervision
( ) 1 needs one person to assist
( ) 0 needs two people to assist or supervise to be safe

6. STANDING UNSUPPORTED WITH EYES CLOSED

INSTRUCTIONS:
Please close your eyes and stand still for 10 seconds.
( ) 4 able to stand 10 seconds safely
( ) 3 able to stand 10 seconds with supervision
( ) 2 able to stand 3 seconds
( ) 1 unable to keep eyes closed 3 seconds but stays safely
( ) 0 needs help to keep from falling

7. STANDING UNSUPPORTED WITH FEET TOGETHER

INSTRUCTIONS:

Place your feet together and stand without holding on.

( ) 4 able to place feet together independently and stand 1 minute safely
( ) 3 able to place feet together independently and stand 1 minute with supervision
( ) 2 able to place feet together independently but unable to hold for 30 seconds
( ) 1 needs help to attain position but able to stand 15 seconds feet together
( ) 0 needs help to attain position and unable to hold for 15 seconds

Berg Balance Scale continued.....

8. REACHING FORWARD WITH OUTSTRETCHED ARM WHILE STANDING

INSTRUCTIONS:

Lift arm to 90 degrees. Stretch out your fingers and reach forward as far as you can. (Examiner places a ruler at the end of fingertips when arm is at 90 degrees. Fingers should not touch the ruler while reaching forward. The recorded measure is the distance forward that the fingers reach while the subject is in the most forward lean position. When possible, ask subject to use both arms when reaching to avoid rotation of the trunk.)

( ) 4 can reach forward confidently 25 cm (10 inches)
( ) 3 can reach forward 12 cm (5 inches)
( ) 2 can reach forward 5 cm (2 inches)
( ) 1 reaches forward but needs supervision
( ) 0 loses balance while trying/requires external support

9. PICK UP OBJECT FROM THE FLOOR FROM A STANDING POSITION

INSTRUCTIONS:

Pick up the shoe/slipper, which is place in front of your feet.

( ) 4 able to pick up slipper safely and easily
( ) 3 able to pick up slipper but needs supervision
( ) 2 unable to pick up but reaches 2-5 cm(1-2 inches) from slipper and keeps balance independently
( ) 1 unable to pick up and needs supervision while trying
( ) 0 unable to try/needs assist to keep from losing balance or falling

10. TURNING TO LOOK BEHIND OVER LEFT AND RIGHT SHOULDERS WHILE STANDING

INSTRUCTIONS:

Turn to look directly behind you over toward the left shoulder. Repeat to the right. Examiner may pick an object to look at directly behind the subject to encourage a better twist turn.

( ) 4 looks behind from both sides and weight shifts well
( ) 3 looks behind one side only other side shows less weight shift
( ) 2 turns sideways only but maintains balance
( ) 1 needs supervision when turning
( ) 0 needs assist to keep from losing balance or falling

11. TURN 360 DEGREES

INSTRUCTIONS:

Turn completely around in a full circle. Pause. Then turn a full circle in the other direction.

( ) 4 able to turn 360 degrees safely in 4 seconds or less
( ) 3 able to turn 360 degrees safely one side only 4 seconds or less
( ) 2 able to turn 360 degrees safely but slowly
( ) 1 needs close supervision or verbal cuing
( ) 0 needs assistance while turning

12. PLACE ALTERNATE FOOT ON STEP OR STOOL WHILE STANDING UNSUPPORTED

INSTRUCTIONS:

Place each foot alternately on the step/stool. Continue until each foot has touch the step/stool four times.

( ) 4 able to stand independently and safely and complete 8 steps in 20 seconds
( ) 3 able to stand independently and complete 8 steps in > 20 seconds
( ) 2 able to complete 4 steps without aid with supervision
( ) 1 able to complete > 2 steps needs minimal assist
( ) 0 needs assistance to keep from falling/ unable to try

13. STANDING UNSUPPORTED ONE FOOT IN FRONT

INSTRUCTIONS: (DEMONSTRATE TO SUBJECT)

Place one foot directly in front of the other. If you feel that you cannot place your foot directly in front, try to step far enough ahead that the heel of your forward foot is ahead of the toes of the other foot. (To score 3 points, the length of the step should exceed the length of the other foot and the width of the stance should approximate the subject's normal stride width.)

( ) 4 able to place foot tandem independently and hold 30 seconds
( ) 3 able to foot ahead independently and hold 30 seconds
( ) 2 able to take small step independently and hold 30 seconds
( ) 1 needs help to step but can hold 15 seconds
( ) 0 loses balance while stepping or standing

14. STANDING ON ONE LEG

INSTRUCTIONS:

Stand on one leg as long as you can without holding on.

( ) 4 able to lift leg independently and hold > 10 seconds
( ) 3 able to lift leg independently and hold 5-10 seconds
( ) 2 able to lift leg independently and hold ≥ 3 seconds
( ) 1 tries to lift leg unable to hold 3 seconds but remains standing independently.
( ) 0 unable to try of needs assist to prevent fall

Tinetti Performance Oriented Mobility Assessment (POMA)

Description:

The Tinetti assessment tool is an easily administered task-oriented test that measures an older adult's gait and balance abilities.

Equipment needed:

- Hard armless chair
- Stopwatch or wristwatch
- 15 ft walkway

Completion Time:

10-15 minutes

Scoring:

A three-point ordinal scale, ranging from 0-2 where "0" indicates the highest level of impairment and "2" the individual's independence.

Total Balance Score = 16
Total Gait Score = 12
Total Test Score = 28

Interpretation:

25-28 = low fall risk
19-24 = medium fall risk
< 19 = high fall risk

Tinetti ME. Performance-oriented assessment of mobility problems in elderly patients. JAGS 1986; 34: 119-126. (Scoring description: PT Bulletin Feb. 10, 1993)

Tinetti Performance Oriented Mobility Assessment (POMA)

Balance Tests
Initial instructions: Subject is seated in hard, armless chair. The following manoeuvres are tested.

1. Sitting Balance

Leans or slides in chair	=0
Steady, safe	=1

2. Arises

Unable without help	=0
Able, uses arms to help	=1
Able without using arms	=2

3. Attempts to Arise

Unable without help	=0
Able, requires > 1 attempt	=1
Able to rise, 1 attempt	=2

4. Immediate Standing Balance (first 5 seconds)

Unsteady (swaggers, moves feet, trunk sway)	=0
Steady but uses walker or other support	=1
Steady without walker or other support	=2

5. Standing Balance

Unsteady	=0
Steady but wide stance(medial heals > 4 inches apart) and uses cane or other support	=1
Narrow stance without support	=2

6. Nudged
(subject at maximum position with feet as close together as possible, examiner pushes lightly on subject's sternum with palm of hand 3 times)

Begins to fall	=0
Staggers, grabs, catches self	=1
Steady	=2

7. Eyes Closed (at maximum position of item 6)

Unsteady	=0
Steady	=1

8. Turing 360 Degrees

Discontinuous steps	=0
Continuous steps	=1
Unsteady (grabs, staggers)	=0
Steady	=1

9. Sitting Down

Unsafe (misjudged distance, falls into chair)	=0
Uses arms or not a smooth motion	=1
Safe, smooth motion	=2

BALANCE SCORE: **/16**

Gait Tests

INITIAL INSTRUCTIONS:
Subject stands with examiner, walks down hallway or across room, first at "usual" pace, then back at "rapid, but safe" pace (using usual walking aids)

10. Initiation of Gait (immediately after told to "go"

Any hesitancy or multiple attempts to start	=0
No hesitancy	=1

11. Step Length and Height

Right swing foot	
Does not pass left stance foot with step	=0
Passes left stance foot	=1
Right foot does not clear floor completely	
With step	=0
Right foot completely clears floor	=1
Left swing foot	
Does not pass right stance foot with step	=0
Passes right stance foot	=1
Left foot does not clear floor completely	
With step	=0
Left foot completely clears floor	=1

12. Step Symmetry

Right and left step length not equal (estimate)	=0
Right and left step length appear equal	=1

13. Step Continuity

Stopping or discontinuity between steps	=0
Steps appear continuous	=1

14. Path (estimated in relation to floor tiles, 12-inch diameter; observe excursion of 1 foot over about 10 ft. of the course)

Marked deviation	=0
Mild/moderate deviation (uses walking aid)	=1
Straight without walking aid	=2

15. Trunk

Marked sway or uses walking aid	=0
No sway but flexion of knees or back or	
Spreads arms out while walking	=1
No sway, no flexion, no use of arms, and no	
Use of walking aid	=2

16. Walking Stance

Heels apart	=0
Heels almost touching while walking	=1

GAIT SCORE = /12

TOTAL SCORE (Gait + Balance) = /28

< 19	high fall risk,
19-24	medium fall risk,
25-28	low fall risk

Tinetti Performance Oriented Mobility Assessment (POMA)
Balance Tests: Subject is seated on hard, armless chair
SITTING BALANCE Leans or slides in chair =0, Steady, safe =1
ARISES Unable without help =0; Able, uses arms =1, Able without using arms = 2
ATTEMPTS TO RISE: Unable w/o help=0; Able, requires > 1 attempt =1; Able in 1 attempt =2
IMMEDIATE STANDING BALANCE (first 5 seconds) Unsteady (sway/stagger/feet move)=0; Steady, w/ support =1;Steady w/o support =2
STANDING BALANCE Unsteady =0; Steady, stance > 4 inch BOS & requires support =1; Narrow stance, w/o support =2
STERNAL NUDGE (feet close together) Begins to fall =0; Staggers, grabs, catches self =1; Steady =2
EYES CLOSED (feet close together) Unsteady =0; Steady =1
TURNING 360 DEGREES Discontinuous steps =0; Continuous steps =1
TURNING 360 DEGREES Unsteady (staggers, grabs) =0;Steady =1
SITTING DOWN Unsafe (misjudges distance, falls) =0;Uses arms, or not a smooth motion =1; Safe, smooth motion =2
BALANCE SCORE TOTAL

Tinetti Performance Oriented Mobility Assessment (POMA)
GAIT INITATION (immediate after told "go") Any hesitancy, multiple attempts to start =0; No hesitancy =1
STEP LENGTH R swing foot passes L stance leg =1; L swing foot passes R =1
FOOT CLEARANCE R foot completely clears floor =1; L foot completely clears floor =1
STEP SYMMETRY R and L step length unequal =0; R and L step length equal=1
STEP CONTINUITY Stop/discontinuity between steps =0; Steps appear continuous =1
PATH (excursion) Marked deviation =0; Mild/moderate deviation or use of aid =1; Straight without device=2
TRUNK Marked sway or uses device =0; No sway but knee or trunk flexion or spread arms while walking =1; None of the above deviations=2
BASE OF SUPPORT Heels apart =0; Heels close while walking =1
GAIT SCORE TOTAL
ASSISTIVE DEVICE
TOTAL SCORE (BALANCE + GAIT) FALL RISK (minimal >23, Mod. 19-23, High < 19)
Therapist initials

Dynamic Gait Index

Description:
Developed to assess the likelihood of falling in older adults. Designed to test eight facets of gait.

Equipment needed:
- Box (Shoebox),
- Cones (2),
- Stairs,
- 20' walkway, 15" wide

Completion:
Time 15 minutes

Scoring:
A four-point ordinal scale, ranging from 0-3. "0" indicates the lowest level of function and "3" the highest level of function. Total Score = 24

Interpretation:
< 19/24 = predictive of falls in the elderly
> 22/24 = safe ambulators

1. Gait level surface
INSTRUCTIONS:
Walk at your normal speed from here to the next mark (20')
Grading: Mark the lowest category that applies.
(3) Normal: Walks 20', no assistive devices, good sped, no evidence for imbalance, normal gait pattern
(2) Mild Impairment: Walks 20', uses assistive devices, slower speed, mild gait deviations.
(1) Moderate Impairment: Walks 20', slow speed, abnormal gait pattern, evidence for imbalance.
(0) Severe Impairment: Cannot walk 20' without assistance, severe gait deviations or imbalance.

2. Change in gait speed
INSTRUCTIONS:
Begin walking at your normal pace (for 5'), when I tell you "go," walk as fast as you can (for 5'). When I tell you "slow," walk as slowly as you can (for 5').
Grading: Mark the lowest category that applies.
(3) Normal: Able to smoothly change walking speed without loss of balance or gait deviation. Shows a significant difference in walking speeds between normal, fast and slow speeds.
(2) Mild Impairment: Is able to change speed but demonstrates mild gait deviations, or not gait deviations but unable to achieve a significant change in velocity, or uses an assistive device.
(1) Moderate Impairment: Makes only minor adjustments to walking speed, or accomplishes a change in speed with significant gait deviations, or changes speed but has significant gait deviations, or changes speed but loses balance but is able to recover and continue walking.
(0) Severe Impairment: Cannot change speeds, or loses balance and has to reach for wall or be caught.

3. Gait with horizontal head turns
INSTRUCTIONS:
Begin walking at your normal pace. When I tell you to "look right," keep walking straight, but turn your head to the right. Keep looking to the right until I tell you, "look left," then keep walking straight and turn your head to the left. Keep your head to the left until I tell you "look straight," then keep walking straight, but return your head to the center.
Grading: Mark the lowest category that applies.
(3) Normal: Performs head turns smoothly with no change in gait.
(2) Mild Impairment: Performs head turns smoothly with slight change in gait velocity, i.e., minor disruption to smooth gait path or uses walking aid.
(1) Moderate Impairment: Performs head turns with moderate change in gait velocity, slows down, staggers but recovers, can continue to walk.
(0) Severe Impairment: Performs task with severe disruption of gait, i.e., staggers outside 15" path, loses balance, stops, reaches for wall.

4. Gait with vertical head turns
INSTRUCTIONS:
Begin walking at your normal pace. When I tell you to "look up," keep walking straight, but tip your head up. Keep looking up until I tell you, "look down," then keep walking straight and tip your head down. Keep your head down until I tell you "look straight," then keep walking straight, but return your head to the center.
Grading: Mark the lowest category that applies.
(3) Normal: Performs head turns smoothly with no change in gait.
(2) Mild Impairment: Performs head turns smoothly with slight change in gait velocity, i.e., minor disruption to smooth gait path or uses walking aid.
(1) Moderate Impairment: Performs head turns with moderate change in gait velocity, slows down, staggers but recovers, can continue to walk.
(0) Severe Impairment: Performs task with severe disruption of gait, i.e., staggers outside 15" path, loses balance, stops, reaches for wall.

5. Gait and pivot turn
INSTRUCTIONS:
Begin walking at your normal pace. When I tell you, "turn and stop," turn as quickly as you can to face the opposite direction and stop.
Grading: Mark the lowest category that applies.
(3) Normal: Pivot turns safely within 3 seconds and stops quickly with no loss of balance.
(2) Mild Impairment: Pivot turns safely in > 3 seconds and stops with no loss of balance.
(1) Moderate Impairment: Turns slowly, requires verbal cueing, requires several small steps to catch balance following turn and stop.
(0) Severe Impairment: Cannot turn safely, requires assistance to turn and stop.

6. Step over obstacle
INSTRUCTIONS:
Begin walking at your normal speed. When you come to the shoebox, step over it, not around it, and keep walking.
Grading: Mark the lowest category that applies.
(3) Normal: Is able to step over the box without changing gait speed, no evidence of imbalance.
(2) Mild Impairment: Is able to step over box, but must slow down and adjust steps to clear box safely.

(1) Moderate Impairment: Is able to step over box but must stop, then step over. May require verbal cueing.
(0) Severe Impairment: Cannot perform without assistance.

7. Step around obstacles
INSTRUCTIONS:
Begin walking at normal speed. When you come to the first cone (about 6' away), walk around the right side of it. When you come to the second cone (6' past first cone), walk around it to the left.
Grading: Mark the lowest category that applies.
(3) Normal: Is able to walk around cones safely without changing gait speed; no evidence of imbalance.
(2) Mild Impairment: Is able to step around both cones, but must slow down and adjust steps to clear cones.
(1) Moderate Impairment: Is able to clear cones but must significantly slow, speed to accomplish task, or requires verbal cueing.
(0) Severe Impairment: Unable to clear cones, walks into one or both cones, or requires physical assistance.

8. Steps
INSTRUCTIONS:
Walk up these stairs as you would at home, i.e., using the railing if necessary. At the top, turn around and walk down.
Grading: Mark the lowest category that applies.
(3) Normal: Alternating feet, no rail.
(2) Mild Impairment: Alternating feet, must use rail.
(1) Moderate Impairment: Two feet to a stair, must use rail.
(0) Severe Impairment: Cannot do safely.

TOTAL SCORE: _____ / 24

Reference:
Herdman SJ. Vestibular Rehabilitation. 2nd ed. Philadelphia, PA: F.A.Davis Co; 2000.
Shumway-Cook A, Woollacott M. Motor Control Theory and Applications, Williams and Wilkins Baltimore, 1995: 323-32

Medical Research Council Dyspnoea Scale [5] for grading the degree of a patient's breathlessness

1. Not troubled by breathlessness except on strenuous exercise
2. Short of breath when hurrying or walking up a slight hill
3. Walks slower than contemporaries on the level because of breathlessness, or has to stop for breath when walking at own pace
4. Stops for breath after about 100 m or after a few minutes on the level
5. Too breathless to leave the house, or breathless when dressing or undressing

Modified Borg Dyspnoea Scale [6]

Patient Instructions for Borg Dyspnoea Scale

"This is a scale that asks you to rate the difficulty of your breathing. It starts at number 0 where your breathing is causing you no difficulty at all and progresses through to number 10 where your breathing difficulty is maximal. How much difficulty is your breathing causing you right now?"

0	Nothing at all
0.5	Very, very slight (just noticeable)
1	Very slight
2	Slight
3	Moderate
4	Somewhat severe
5	Severe
6	
7	Very severe
8	
9	Very, very severe (almost maximal)
10	Maximal

[5] http://www.nice.org.uk/usingguidance/commissioningguides/pulmonaryrehabilitationserviceforpatientswithcopd/specifyingapulmonaryrehabilitationserviceforpatientswithcopd/mrcdyspnoeascale/mrc_dyspnoea_scale.jsp

[6] http://www.pulmonaryrehab.com.au/pdfs/resourcesPatient%20Assessment_BorgScale.doc

Appendix Three: Case Studies

Case Study 1

Examination [7]

History and Systems Review: Ms. J was a 50-year woman, referred to outpatient physical therapy because of mild clumsiness and balance problems. Although she had not fallen, her physician felt she was at increased risk for falling. Ms. J had not been formally diagnosed with Huntington's disease. However, her father died from complications of the disease five years previously. She chose not to be tested for the Huntington gene mutation. Through MRI examination, mild degeneration of the caudate and putamen was identified. Past medical history included treatment for depression. The physician felt that there was sufficient cause to establish suspicion of HD. Ms. J was an active woman and avid bike rider. She regularly rode to work a distance of about 7 miles and had participated in many bike adventure trips. She was employed by a large company as an administrative assistant. She was married and had a 15-year son at the time of referral.

[7] Reproduced with permission from Smith, M, Danoff, J, Jain, M & Long, T. (2007) Genetic Disorders: Implications for Allied Health Professionals: Two Case Studies. International Journal of Allied Health Sciences and Practice. http://ijahsp.nova.edu. Vol 5, No 4 ISSN 1540-580X

Test and Measures

Aerobic capacity and endurance: This patient's resting heart rate was 80 bpm, with blood pressure of 100/70.

Arousal, attention and cognition: Ms. J was oriented to person, place and time. She did not feel that her memory or attention had diminished, nor did her family report any personality changes.

Sensation, integrity and reflex integrity: Sensation to light touch, pinprick and proprioception was intact. Deep tendon reflexes were brisk without clonus.

Motor function: Ms. J demonstrated mild, low amplitude, increasing to high amplitude (under stress), choreiform movement. Finger to thumb tapping and diadochokinesia was arrhythmic and slow. She displayed mild bilateral in coordination with finger-to-nose and finger-to-finger tests. Her ability to bike to work was only minimally affected at the time of referral. She rode slowly and cautiously. Some days she had to convince herself to bike because she felt so tired.

Range of motion and muscle performance: Passive and active range of motion were within normal limits. Muscle strength was generally 4/5.

Gait, locomotion, and balance: This patient could sit independently without any back support for an indefinite period of time. She could stand up from a sitting position without assistance, but slowly. She stood independently without support. Although she could stand with her feet together, her gait was wide-based. Walking on a flat surface, ascending and descending stairs, and on uneven terrain were all within normal limits. She had difficulty with tandem walking and heel-to-toe walking.

Self-care and home management: She planed and made all family meals and completed all household tasks. Ms. J complained that she was slower performing these tasks, and she felt disorganized. She was independent with all her self help skills. Her home was a large two-story house where most of the flooring was wood with a variety of floor coverings.

Evaluation, Diagnosis, Prognosis

Ms. J scored 11/13 on the Huntington's Disease Functional Capacity Scale. She did not report difficulty performing her job related tasks. She did require some assistance with domestic and financial affairs due to her lack of organization. She was living at home and was independent with all self care and home management.

Due to the choreiform movements, Ms. J was beginning to experience difficulty with driving, especially at night. This impacted her ability to drive her son to his sporting events. She had been actively involved in organizing fundraisers, practices and other team related events for her son, but was considering dropping these activities.

At the time of this writing Ms. J's deficits were very minimal. According to the Huntington's Disease Functional Capacity Scale, she was at Stage 1 of the condition. She was independent with all of her ADL's. She was continuing her work as a secretary and was able to carry out her household responsibilities with minimal assistance. On some days she did not feel comfortable biking or driving to work, so she would take the bus, which required additional planning. She was especially concerned about being able to maintain her biking ability.

Our therapeutic goals for this individual focused on maintaining her ability to complete all ADL's independently, preservation of present range of motion and strength, and overall preservation of her ability to move independently. To accomplish these goals, we recommended an exercise program to help maintain her range of motion and strength. In addition, anticipatory guidance was provided to her and her family about the consequences of HD and the need to plan for eventual movement limitations. She was instructed in relaxation strategies to decrease her stress and minimize her chorea movements. We also recommended that she take mini-breaks at work, which we believe would help her to stay focused and organized.

Intervention

Due to her status, ongoing intervention was not currently necessary. One to two sessions of education and anticipatory guidance were considered adequate as Ms. J and her family were knowledgeable about the consequences of the condition. Ms. J was encouraged to become involved in a 3-5 times per week community-based exercise program. This program consisted of a variety of aerobic activities such as stationary bike, treadmill, elliptical training, and aerobic exercise classes. Additionally, a comprehensive strength training program was designed. To maintain flexibility, Ms. J chose to participate in a flexibility and strength class and planned also to participate in a yoga program. Because biking was of particular concern to Ms. J, she engaged a trainer who was knowledgeable in biking exercises.

A variety of referral sources that she might benefit from over the course of the next 6 months were presented to Ms. J. These were intended to help her maintain an organized environment at home, continue with the performance of her household duties, and structure management of her financial affairs. The referrals included an organization specialist, a HD support group, a physical therapist who specialized in office ergonomics, and an occupational therapist who specialized in assistive technology.

Re-examination and Goal Modification

The median survival after diagnosis of Huntington's disease is approximately 21 years; therefore, re-examination and follow-up for supportive needs may be needed.[23] Because Ms. J indicated that she would like to maintain an active role in decision making regarding her condition for as long as possible, she will need to establish collaborative relationships with a variety of professionals. A 6 month re-examination with the physical therapist is appropriate. In addition to re-examining physical status and maintaining functional status, the therapist and Ms. J would be expected to discuss the need for any additional resources.

Understanding the progressive nature of the condition is imperative for the family to prepare themselves for future situations, physically and psychologically. As Ms. J's condition deteriorates, physical therapy will modulate from a consultative role to a direct service provider. As the limitations become more severe, physical therapy goals will evolve from prevention to restoration to maintenance – with an increased emphasis on management of impairments. Comprehensive care for Ms. J would entail collaboration among a variety of professionals. Occupational therapy will be needed to address self-care issues; and speech pathology, to address communication, feeding and swallowing issues. Having the family maintain a link to rehabilitation services as the condition progresses may minimize feelings of isolation.

Outcome

This patient was provided with a variety of strategies to maintain safe ambulation, strength, and flexibility. Over the next decade, it is probable that she will become increasingly dependent on others for assistance with activities of daily living, home management, and professional responsibilities. Future issues for her will include dependence related to ADL's, domestic management, and inability to work. Prior to her inability to continue working, her employer may need to make adaptations to her work environment in order to comply with ADA regulations. The adaptations may include changing her workstation, providing her with a time management device, and project management software to help her stay organized. Although she had chosen not to get tested for the genetic mutation, we believe she should consider having her son tested. We have encouraged her to meet with a genetics counselor to discuss this. Ms. J's decision to avoid genetic testing was based on the knowledge that there is currently no cure or specific effective treatment for HD, and she would not be able to change the eventuality of having HD. However, when her son reaches the age of maturity, he may choose to be tested. Knowing that he has the genetic mutation may help him to prepare for its eventual presentation. Unfortunately, this information also may lead to stigmatization and discrimination. There have been cases in which both health and life insurers have denied coverage to those with a genetic disease although the individuals were currently healthy.[31] Additionally, when Ms. J's son enters the workforce, he could experience workplace discrimination.[31,32,33] Because there is an increased risk for suicide in this population[21], psychological counseling is an important service to which Ms. J's son may desire access. Thus, any health insurance should include behavioral care.

Case Study 2

Application of exercise prescription principles: a case report [8]

HD is a degenerative disease of the basal ganglia with a major impact on mobility, cognition and behaviour over 15-20 years. People with HD have been shown to be weaker (Busse et al. 2008), walk less and fall more (Busse et al. 2009) than an age-matched control group. Evidence from exercise studies in animals (Zuccato & Cattaneo 2007), healthy people (Winter et al. 2007) as well as individuals with other neurological conditions (White & Castellano 2008) suggest that exercise interventions have potential to offer health, social, cognitive and coordination benefits for individuals with HD. To date, there is no reporting of the progression and effect of controlled exercise prescription for people with HD, although exercise is usually considered to be one component of multi-disciplinary rehabilitation for people with HD (Zinzi et al. 2007).

Adverse effects, namely exercise-induced muscle fatigue, pain, elevated creatine kinase level, and worsening of his running performance have however been reported in one semi-professional athlete at risk for HD when training for a marathon (Kosinski 2007). In this single case, excessive training is suggested to have resulted in myopathy in HD before the appearance of other neurological symptoms. Investigation of the exercise response in this condition is therefore indicated. We examined the feasibility and effect of delivering aerobic and anaerobic exercise in an individual with this complex condition.

A 39 year old female, weight (74 kg); height (157 cm), with genetically confirmed mid-stage HD, completed a 7 week long supervised exercise programme. She demonstrated minimal chorea and no marked behavioral or cognitive impairment, was living in her own home but

[8] Reproduced with permission from ME Busse1 H Dawes2,5, A Meaney2 and AE Rosser 3;4

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5 Department of Clinical Neurology, University of Oxford

requiring moderate assistance with activities of daily living. Informed consent was obtained. Outcome assessment was conducted by an independent physiotherapist at baseline and after 8 weeks. Balance and functional mobility (Berg balance scale (BBS) and Timed Up and Go (TUG)) (Wade 1992), maximum voluntary contraction of the knee extensor and flexor muscle groups (Powertrak Hand-held dynamometer; JTech Medical Utah, USA) self reported falls in the previous 30 days and Physical Activity (International Physical Activity Questionnaire-Long Form (IPAQ-LF) was assessed.

The exercise intervention consisted of aerobic and anaerobic training delivered in a community gym twice a week for seven weeks. Aerobic training was performed with the participant seated in a recumbent cycle (LifeFitness 95 Ri, Illinois, USA). The participant was instructed to cycle so that heart rate (HR) was maintained within an aerobic training zone (55-85 percentage age predicted maximal heart rate % APMHR) (ACSM 2002). Once 20 minutes continuous exercise duration was attained, duration was gradually increased up to 40 minutes. Resistance was manipulated to maintain HR within an aerobic zone. Rating of perceived exertion (RPE, CR10 scale) and HR were recorded (Dawes 2007). Anaerobic strength training was performed at an initial resistance so that 10 repetitions could be performed. Repetitions were progressed, until 2 full sets of 10 could be performed at a given resistance with a 2 minute rest between sets. Resistance was then increased with a resultant decrease in repetitions and the protocol progression then repeated. Leg extension was performed on a seated leg press (LifeFitness). Sit to stand exercises were performed from a standard chair. Functional core stability was trained during anaerobic upperlimb training with exercises [double arm pull-down, a double arm lateral raise, single arm rotation (both sides)] initially performed seated but progressed to standing after 5 sessions using a Dual Cable Crossover resistance machine (LifeFitness). Work done per session is reported by multiplying the weight resistance by the number of completed sets. The resistance for sit to stand was calculated from body weight.

The participant attended all sessions with no adverse symptoms reported or noted. Valid initial assessment of fitness was not possible due to an inability to attain independent cycling cadence for an adequate duration. RPE rating was variable and at variance with that expected for a given HR (Dawes 2007). Low aerobic exercise intensity was achieved; %APHRM (mean ± SD, 64 ±4%). Increasing the duration of aerobic sessions beyond 20 minutes continuous cycling was challenging (mean ± SD, 33 ± 8 mins).

Maximal achievable pedaling cadence progressed from 55 to 101rpm (mean ± SD 73 ± 20rpm). Anaerobic exercises were steadily progressed for all muscle groups. Change in volume of work done (kg) from the second session to the final session was as follows: leg press 2075kg, sit to stand 740kg, trapesius 62.5kg, latissimus dorsi 62.5 kg, transversus lumborum 62.5kg. Change in balance, strength and mobility over the seven weeks are reported in Table 1.

Table 1 Functional outcomes

Measure	Assessment 1	Assessment 2
Timed up and go (sec)	17.9	13.2
Berg Balance Scale	18/56	44/56
Knee extensor strength R (N)	123	160
Knee extensor strength L (N)	147	151
Knee flexor strength R (N)	114	121
Knee flexor strength L (N)	98	99
Reported falls (number in previous 30 days)	3	0
Participation in physical activity/week (mins) (IPAQ-LF)	396	1952
Time sitting/week (mins) (IPAQ-LF)	3360	900

We have demonstrated that it is feasible to deliver a supervised progressive training programme that includes both aerobic and anaerobic components in an individual with HD. Aerobic training intensities were achieved throughout, but increasing training duration beyond 20 minutes duration was challenging. RPE was not a valid intensity measure and its use requires further examination in people with HD. In this single case, improvements in strength, balance, self reported physical activity and self reported falls occurrence and no adverse side effects were observed. Our findings are encouraging and support further investigation into therapeutic exercise interventions for people with HD.

Appendix Four: Patient and client education, suggestions for general physical conditioning activities

The following are some exercises that can be done to improve general conditioning and physical fitness. Ideally, cardiovascular exercises should be done for at least 20 minutes a day, 3 a week. It is important to maintain and potentially increase caloric intake after starting any form of exercise. The person with HD should discuss any exercise programme with their doctor before starting out.

Walking is a good way to promote cardiovascular health. It is easy to do and can be done anywhere. Some places that are good to walk are on a track, around the block, or around the mall. Start with the person with HD walking for **10 minutes a day 3 times a week**, slowly at first, and gradually progressing by increasing walking speed/pace. Walking pace should be comfortable (Use the rate of perceived exertion scale (RPE) to help determine intensity level of exercise).

Eventually, walking should be fast enough to build up a sweat but slow enough to not be out of breath (if walking with a companion, the person should be able to carry on a conversation with him or her). After completion of the walk, the person with HD should walk slowly for at least 1 or 2 minutes to cool down and not stop abruptly. Once the person with HD is comfortable walking for 10 minutes a day, they can try to increase the time to 15 minutes and finally to 20 minutes. They should be advised that if they feel short of breath or cannot breathe, to **stop immediately,** rest and see their doctor.

Using an exercise bike is another good way to improve cardiovascular health. If the person with HD does not have a bike at home, they could consider using one at a local gym. Once again, the goal is to reach 20 minutes of biking 3 times a week. Start out by doing 10 minutes of biking and then gradually increase the amount of time to 20 minutes. As with walking, starting off slowly, and gradually pick up the pace, whilst choosing a comfortable resistance is essential.

Exercising in the pool is also a good way to achieve cardiovascular training, as well as improve coordination and balance. To start with, the person with HD might want to try some **exercises** in the pool; for example standing in the water and kicking legs, one at a time, out to the sides. This can also be done with the arms. If able, the person with HD can **swim** a few laps around the pool. If they have difficulty swimming, they could try using a kick board, which supports the upper body. If your local gym has any organized pool classes, such as Aqua-aerobics, or programs offered for people with disabilities, the person with HD might consider enrolling in them but be sure to enquire about the exact nature of the exercises and whether they are appropriate for people with HD. Poolside supervision is important for safety.

Appendix Five: Frequently asked questions

Q1 – Can someone with HD manage with a walking aid or electric wheelchair?

A – This is very dependent on the individual; cognitively, people with HD struggle with divided attention and may not have the co-ordination or concentration to use a walking aid. Physically, people can struggle with walking aids if their chorea is severe meaning that they can't place the stick or frame and tend to pick them up instead.

Walking sticks: In the early stages of the disease some people like to use a walking stick; this provides support and can also to highlight the fact that they have a disability and are not drunk. If people are struggling with the placement of the walking stick weights in the bottom can be useful.

Walking frames: Those who do use a frame tend to do better with a frame with wheels as this is easier to co-ordinate than a standard frame. Some people find they can manage these when they are on a flat surface but can't cope with steps up to the pavement etc. Very few people manage an electric wheelchair or scooter due to the need for divided attention. Those who do use one tend to need constant supervision and a large and clear area.

They feel more secure and calm with a person next to them holding their arm. Some people relax with a person next to them, so that they can walk and perform better.

Q2 – My client is falling from their chair when they are trying to get up for a cigarette, but we can't constantly supervise him. What can we do?

A – These issues often arise in the mid stages of HD where someone is still mobile but falling, they are focusing on the thing they want (here the cigarette) and are often unaware of the risks. These cases require good behavioural management and routine e.g. agreeing on half hourly cigarettes when there can be a carer there to supervise.

This would also be a good time to discuss protective clothing with your client such as knee/elbow pads and helmets.

Q3 – My client is kicking out due to spasms in her legs when she is using the shower chair.

A – This could be behavioural rather than physical: Is the person kicking out as it is their only way of communicating the fact that they don't like the situation they are in? Consideralking to your client to see if you can establish whether they are unhappy in the shower (if communication is difficult consider involving the Speech and Language Therapist). Looking into how the person is being showered, often people are happier with one carer rather than another and it is worth considering what they are doing differently.

People with HD often don't like surprises and need regular re-assurance. It would be useful to get into a good routine where you are prepared for the shower before starting, and guide the person with what's happening next, how long you have got left, etc. It could also be that the at this stage, a shower chair is no longer suitable due to decreased trunk control, and possibly a shower trolley may be more suitable.
In terms of equipment it will be worth considering if the chair is still suitable – do they need a specialist shower chair such as 'tilt in space' or would they be more comfortable using a shower trolley?

Q4 – My client has severe chorea; he is getting trapped and hurting himself on the cotsides

A – People with severe chorea may find themselves in all sorts of positions in the bed and can get trapped in standard cot sides. Specialist equipment can help e.g. inflatable cot sides that surround the bed and link to a sheet under the mattress meaning that the person can't get trapped. Medications are available for chorea but the side effects should always be considered – this requires discussion with the neurologist.

The client could also be anxious about something whilst in bed. Anxiety may increase the chorea. Soothing music or a weighted blanket may help. If entrapment is a real danger and the bed cannot be lowered to the floor, it could be safer to put the mattress on the floor. Relaxation exercises, massage, and calming behaviour of carers may also be helpful.

Q5 – My client can no longer stand and transfer but is going into spasm when using a hoist, how can we transfer him safely?

A – First it is important to make sure that the hoist is being used properly, anti-spasm slings are available and some manufacturers will make bespoke slings considering your client's needs. Carers often find that ceiling hoists are more appropriate for people with HD as they are fixed.

Before the transfer, the patient should be well informed and calmed down in order to optimize the transfer. Shortage of time or other stress might be something that the HD patient senses and reacts to which can evoke dystonia/spasm.

Resources

The following is a listing of web-based and printed resources available to assist those with Huntington's Disease, their families and their caretakers in learning more about and coping with the disease process. This is by no means a comprehensive list, but it is a good springboard from which to find additional information. The views expressed on websites created by individuals reflect only their views and not the specific views of EHDN.

Web Resources

Advanced Stages of Huntington's Disease Caregivers Handbook http://huntingtondisease.tripod.com/advancedstagesofhd/

Physiotherapy and Occupational Therapy for Huntington's Disease http://huntingtondisease.tripod.com/therapy/

Advanced Stages of Huntington's Disease Caregivers Handbook – Exercise and Fitness http://huntingtondisease.tripod.com/advancedstagesofhd/id8.html

Caring for People with Huntington's Disease http://www.kumc.edu/hospital/huntingtons/

Disabled Living Foundation. Choosing a chair and chair accessories: http://www.dlf.org.uk/factsheets/pdf/Choosing_a_chair_and_chair_accessories_sponsored.pdf

European Huntington's Disease Network https://www.euro-hd.net/html/network

Family Caregiver Alliance http://www.caregiver.org/caregiver/jsp/content_node.jsp?nodeid=574

Hereditary Disease Foundation http://www.hdfoundation.org/home.php

HD Information Site – compiled by a librarian with an interest in HD http://www2.lib.uchicago.edu/~rd13/hd/

The HD Lighthouse http://hdlighthouse.org/see/index.html

An interview with Stewart Blatt, Physical Therapist http://hdlighthouse.org/treatment-care/care/hdltriad/exercise/updates/0045balance.php

Exercise has a more powerful impact on the brain than previously thought http://hdlighthouse.org/treatment-care/care/hdltriad/exercise/updates/0049exercise.php

Huntington's Disease Advocacy Center http://www.hdac.org/

Huntington's Disease Drug Works http://hddrugworks.org/

Off the Couch: A Great Resolution for the New Year http://hddrugworks.org/index.php?option=com_content&task=view&id=243&Itemid=30

Exercise and Huntington's Disease http://hddrugworks.org/index.php?option=com_content&task=view&id=200

Huntington's Disease Society of America www.hdsa.org

Physiotherapy and Occupational Therapy for Huntington's Disease http://www.hdsa.org/images/content/1/1/11286.pdf

Balance, Falls, and Safety in the Home (PowerPoint Presentation) http://www.hdsa.org/images/content/1/1/11423.pdf

Huntington Society of Canada http://www.hsc-ca.org/

Exercise and Fitness http://www.huntingtonsociety.ca/english/uploads/Exercise_and_Fitness(1).pdf

Preventing Falls http://www.huntingtonsociety.ca/english/uploads/Preventing_Falls(1).pdf

The movement disorder http://www.huntingtonsociety.ca/english/uploads/The_Movement_Disorder(1).pdf

Huntington Study Group http://www.huntington-study-group.org/

Huntington's Working Group: Promoting Excellence in End of Life Care http://www.promotingexcellence.org/i4a/pages/Index.cfm?pageID=3690

International Huntington Association http://www.huntington-assoc.com/

Physical Therapy Intervention for People with Huntington's Disease http://www.huntington-assoc.com/physical.pdf

National Human Genome Research Institute http://www.genome.gov/10001215

Neurology Channel—HD http://www.neurologychannel.com/huntingtons/index.shtml

Taking Care of YOU: Self-Care for Family Caregivers http://www.caregiver.org/caregiver/jsp/content_node.jsp?nodeid=847

The Huntington's Disease Association http://www.hda.org.uk/shop/shop.php

The Huntington's Disease Outreach Project for Education: Stanford University http://www.stanford.edu/group/hopes/

Print Resources

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*Order Form for books/resources from the Huntington's disease Association: http://www.hda.org.uk/shop/HDA-Order-Form.pdf

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