



EUROPEAN HUNTINGTON'S DISEASE NETWORK

# *Occupational Therapy for People with Huntington's Disease: Best Practice Guidelines*



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(with support from the European Huntington's Disease Network  
Standards of Care Occupational Therapists Working Group)**

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## **Part 1**

### **The process of developing the guidelines**

#### **i. Introduction**

Care of people with Huntington's disease varies widely between countries and in different areas in those countries. Resources are not always available to fund and train appropriate staff. None the less, there are many Huntington's disease management clinics worldwide. Some follow guidelines for care provision that they themselves have devised, others react in an ad hoc manner to people with Huntington's disease who are seen in the same clinic as those with a variety of other neurological conditions.

The European Huntington's Disease Network (EHDN) was formed in 2003, by like-minded clinicians and scientists who set out to provide a platform for clinicians, scientists and families to work together to find a cure for Huntington's disease. Working groups were created to provide a forum for specialists to meet and work together in various themes. One such group was the Standards of Care working group, which has specialist groups within it such as the Occupational Therapy working group.

The standards of care working group has produced a consensus view for management of Huntington's disease which reflects the views of highly specialist occupational therapists in their field, and will enable others to provide best care for the person with Huntington's disease and their family. There is little peer-reviewed evidence for our statements. The literature has been reviewed, and it is clear that much work must be undertaken in order to clarify the pathophysiology of some of the features we see in practice, as well as the best management of these phenomena. We expect these guidelines to evolve as evidence emerges and this document to be reviewed and updated accordingly.

#### **ii. The aim of the guideline and target audience**

These guidelines have been developed as a reference guide for occupational therapists working across Europe in a variety of health, social care and speciality settings. A systematic review was conducted to investigate and gain consensus on the range of occupational therapy interventions being used with this population and to establish their effectiveness in ameliorating the impact of Huntington's disease on function. Many of the recommendations in this guideline are based on highly specialist occupational therapy opinion from the EHDN Standards of Care occupational therapists group which has representation from therapists across Europe and the United States of America. It aims to describe and inform contemporary best practice in occupational therapy for people with Huntington's disease.

The aims of these guidelines are:

1. To place the person with Huntington's disease and their family at the centre of all occupational therapy interventions.
2. To support occupational therapists in the assessment and treatment of people with Huntington's disease.
3. To provide a comprehensive overview of currently agreed best practice in Europe.

### **iii. Ratification process**

The guidelines were commissioned by the EHDN and developed as part of the Standards of Care working group which have produced similar guidelines for physiotherapy, speech and language therapy, dietetics and dentistry.

The two initial meetings included occupational therapists from the UK, Sweden and the USA. These meetings focussed on similarities in practice across these countries and discussion of areas to include within the guidelines. The guidelines were written by the main authors and then circulated via e-mail to the other group members for comment and revision. A draft of the guidelines was taken to the College of Occupational Therapists in the UK for ratification.

### **iv. Comments from a Caregiver**

It is important to remember that a person with Huntington's disease has often managed/coped/existed for many years before receiving professional help. Their carers will have adapted their lives as best they can to deal with their difficult situation. Personalities and capabilities vary and all react differently. A person with Huntington's disease will have priorities but not necessarily in the same order as the assessment of a professional. However, their point of view is valid, and if acknowledged, will lead to a better working relationship. Overtime a carer may get exhausted physically, mentally and emotionally. It is important to affirm their role and to learn from them. Sometimes they are reluctant to trust another person with their loved one, but relationships can improve with time. The Huntington's Disease Association (HDA) is a national charity which offers help and advice to those affected by Huntington's disease, whether people with a diagnosis, people at risk, professionals or friends. Regional Care Advisors (RCA) are employed across England and Wales, but the service varies. Some RCAs are welcomed as part of the local multidisciplinary team offering support through different disciplines, whilst others work virtually alone. Current information is available from the HDA website: [www.hda.org.uk](http://www.hda.org.uk)

*(Comments from Jackie Gallop who is a carer and Regional Care Advisor for the HDA in Birmingham, UK)*

## **Background**

### **i. An overview of Huntington's disease**

Huntington's disease is a genetic, chronic neurological condition. The common features of the condition are caused as a result of the gradual deterioration of the caudate nucleus and the putamen found within the basal ganglia.

### **ii. Aetiology**

Huntington's disease is an inherited condition caused by a mutation on chromosome 4 found in gene IT15. The protein which this gene codes is huntingtin, the building blocks of which are the triplet repeat of cytosine, adenine and guanine (CAG). The number of CAG repeats in a normal gene ranges from 15-20. In a person with Huntington's disease the number of CAG repeats are 36 or more. Larger expansions of CAG repeats tend to result in earlier onset of the condition (Stine et al 1993) or a more rapid clinical decline (Ward et al 2006).

Huntington's disease is an autosomal dominant condition, therefore each child of an affected person has a 50% chance of inheriting the Huntington's disease gene and, if they do, they will develop symptoms at some point in their lifetime. There is an equal incidence of the condition in men and women. The gene does not skip a generation consequently, if the parent does not have the gene neither will any of their children.

### **iii. When do symptoms begin?**

The early symptoms of the condition may go undetected for a while and the onset is slow and gradual so it can be difficult for the individual or family member to identify exactly when the symptoms started. Huntington's disease can start at any age but most people develop symptoms between the ages of 35 and 55 years. (Quarrel, 1999).

Juvenile Huntington's disease is a rare disorder comprising of 5-12% of the cases of Huntington's disease. Huntington's disease is difficult to diagnose in children due to the lack of classical features seen in adults. Dementia, reduced movements, rigidity and seizures often dominate the clinical picture in children (Jain 2005).

### **iv. Prevalence**

Current statistics suggest that the prevalence of Huntington's disease is 10 people per 100 000 in UK and Europe. This means that in the UK, which has a population of approximately 55 million, there are about 5500 people with Huntington's disease at any one time. This illustrates that Huntington's disease is a rare disease but if the carers and family members are also counted this condition affects a larger number of people (Quarrel, 1999).

## **v. Diagnosis**

### **a) Genetic counselling and predictive testing**

Genetic counselling is available for people with Huntington's disease, those at risk of developing the condition and any other family members. The aim of genetic counselling is to provide information about Huntington's disease and to discuss specific family situations to enable individuals to make informed choices about their future. Genetic counselling is provided prior to and after predictive testing to discuss the implications of knowing whether or not a person has the affected gene.

The genetic nature of Huntington's disease results in a variety of implications and emotional responses within families.

- Family members may not be aware that Huntington's disease is a genetic condition.
- Family members may not be aware that they are at risk of inheriting the disease.
- Where there is no discernable family history of Huntington's disease individuals may be unaware of the nature and progression of the condition.
- Individuals may not want to know their Huntington's disease status despite showing early symptoms of the condition.

Predictive testing for Huntington's disease has been available since 1993 but many people at risk of inheriting the condition choose not to be tested. Genetic testing can only be undertaken when the individual is over 18 years old and has made this decision following counselling (HDA 2010). However, genetic counselling is available to people before they reach 18. Huntington's disease is often not diagnosed by genetic test but through clinical evaluation once symptoms become problematic.

Individuals who have tested positive for the Huntington's gene and wish to have children can have the foetus tested to see if the gene has been inherited. Preimplantation Genetic Diagnosis (PGD) is available in some areas. This involves the couple having IVF treatment and the embryos being tested for Huntington's disease before being implanted into the woman's womb. Only embryos without the Huntington's disease mutation would be selected for implantation. Advice on foetal testing is available from the HDA fact sheet and local genetics units.

### **b) Clinical evaluation**

Huntington's disease can be diagnosed through a combination of careful history taking and physical testing. The clinician would complete a medical history interview often with a relative of the individual present. This would involve a discussion about any symptoms currently experienced, when they started and any diagnosis or symptoms that a parent or other family members have experienced. The clinician may then complete the motor assessment the scores from which could suggest the presence of Huntington's disease.

**Table 1: Common Signs and Symptoms of Huntington's Disease 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 co-ordination  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, organising, prioritising 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

## vi. Medical treatments

Currently there is no cure for Huntington's disease. Symptoms tend to be treated as they occur or become problematic and there is a range of drugs which can be used for various symptoms. It is important for occupational therapists to be aware of any medications the person with Huntington's disease is on so that they can be aware of any potential side effects that may affect their evaluation or intervention. Any issues relating to drug management should be addressed with the clinician responsible for the medical management of the person.

**Table 2: Main Drug Classes, examples and potential side effects (Quinn and Busse 2009)**

Class of drug	Subclass of drug	Example medications	Potential side effects
Antichoreic	Neuroleptics and atypical antipsychotics	Risperidone (Risperdal) Tiapride (Tiapridex, Synthelabo) Fluphenazine (Prolixin) Olanzapine (Zyprexa) Pimozide (Orap) Geodon (Ziprasidone)	Drowsiness, apathy, extra-pyramidal symptoms, dystonia, akathisia, hypotension, dizziness, headache, insomnia, constipation, dry mouth, weight gain, tardive dyskinesia
	Benzodiazapines	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) Haloperidol (Haldol) Risperidone (Risperdal) Fluphenazine (Prolixin)	Drowsiness, apathy, akathisia, hypotension, dizziness, headache, insomnia, constipation, dry mouth, weight gain, tardive dyskinesia
Antidepressant	Selective Serotonin Re-uptake Inhibitors	Fluoxetine (Prozac) Sertraline (Zoloft) Paroxetine (Paxil) Citalopram (Celexa)	Insomnia, diarrhoea, gastro-intestinal upset, restlessness, weight loss, dry mouth, anxiety, headache
	Tricyclics	Nortriptyline (Pamelor) Amitriptyline (Elavil)	Insomnia, diarrhoea, gastro-intestinal upset, restlessness, weight loss, dry mouth, anxiety, headache
Antiepileptics		Valproate (Depakote) Topiramate (Topomax) Carbamazepine (Tegretol)	Nausea, vomiting, weight gain or loss, cognitive effects, tremor, elevated liver enzymes

### vii. Measuring disability and progression of the condition

Generally, Huntington's disease is a very slowly progressing condition but it can be important to measure change overtime and to measure the impact of treatment interventions. Centres that are involved in the European Huntington's Disease Network Registry project will be completing the Unified Huntington's Disease Rating Scale (UHDRS) annually. This consists of the following sub tests:

- Motor scale (shortened version).
- Cognitive tests: Stroop Test, Symbol Digit Modalities Test, and Verbal Fluency Test.
- Behavioural interview.
- Functional assessment: Total Functional Capacity, Functional Status Scale.

These scales are not particularly relevant to Occupational Therapy so the therapist should select the most appropriate measures they have available. There is a wide range of measures used in occupational therapy but none are specifically designed for an individual with Huntington's disease. Therapists need to consider what it is they want to measure and also to choose measures that are reliable and valid. To date, there is very little research to validate the use of outcome measures in people with Huntington's disease.

Measures that may be useful include:

- Assessment of Motor and Process Skills (AMPS)
- Model of Human Occupation Screening Tool (MoHOST)
- Worker Role Interview (WRI)
- The PRPP (Perceive, Recall, Plan, Perform) functional assessment
- The ADL Taxonomy
- Canadian Occupational Performance Measure (COPM)
- Occupational Performance History Interview version II (OPHI-II) Version 2.1 2004
- Dementia Care Mapping (Assessment in the late stages of the disease)

### **viii) Stages of Huntington's Disease**

There have been several attempts to define the stages of progression of Huntington's disease (Shoulson and Fahn 1979). The Total Functional Capacity Scale divides the disease into 5 stages. However, clinicians tend to describe the stages of disease progression as early, mid and late.

Early disease cannot be defined as the time of diagnosis since diagnosis can be made at variable times. Most individuals present with early neurological and psychiatric features which may have started to cause difficulties at work and home, and many of these individuals are in mid stage disease. As we reach a greater understanding of the early evolution of Huntington's disease, then early stage disease will have a better definition. Recent work has shown that early neuropsychological deficits occur, and physical changes are visible on MRI brain scans. Certainly families report early behaviour and personality changes some years before neurological deficits can be described. Changes may be noticeable at work (depending on the complexity of their job); it may affect driving ability and may become apparent in home tasks which require rapid task switching.

Mid stage disease would be seen to have been reached when the affected person has to cease work, or at least change employment to a post which is less challenging. Executive function decline and some cognitive deficit would be demonstrated. Involuntary movements may be obvious but it is most likely that individuals would still be able to care for themselves.

Late stage disease occurs when employment becomes impossible, the individual is no longer able to live independently, self-care ceases and cognitive decline is obvious. Some individuals maintain the ability to provide limited self-care to a late stage, but the movement disorder, both involuntary and voluntary creates increasing difficulties. Communication may be limited and they will need assistance to participate in social activities.

### **ix) The importance of occupational participation and performance in Huntington's Disease**

As Huntington's disease progresses the individual's ability to participate in and to perform occupations (everyday) tasks becomes increasingly difficult. The type of tasks the individual values and wishes to perform will vary from person to person so it is important for the occupational therapist to spend time with the individual and their family/carers to explore these occupations and clearly identify firstly, what they are, secondly, what the person is finding difficult about the task now and thirdly, what resources are available to assist in occupational participation and performance.

Sullivan et al. (2001) demonstrated that mice with the Huntington's disease gene that were placed in an enriched environment deteriorated at a slower rate than mice with the gene in a non-enriched environment. This would suggest that those people who continue to participate in activity and deal with everyday challenges are likely to reduce the rate of functional decline. More recently, Trembath et al (2010) found age of onset of the illness in human subjects was associated with a more passive lifestyle irrespective of a patient's CAG repeat length. Those with more passive lifestyles experienced onset of the illness 4.6 years earlier than those people who were more active. They conclude being engaged in active pastimes may slow onset of the disease.

Occupational therapists will focus on a range of activities that tend to fall within the following key areas:

- **Self-Care** – activities that enable the individual to survive and to promote and maintain health.
- **Productivity** - includes occupations that make a social or economic contribution or provide economic sustenance.
- **Leisure** – any activity entered into for the purpose of enjoyment such as socialising, creative expressions, outdoor activities and sports. (Law et al 2002).

People have a basic need to participate in activity that has meaning and value to them. Occupations can serve a variety of functions in daily life:

- People develop and maintain function through being active. (Baum 1995)
- Occupations form an important part of each person's social identity and social status, and they influence social development. (Watson and Fourie 2004)
- Occupations contribute to an individual's personal sense of identity. (Hasselkus 2002)
- People create meaning in their lives through what they do. (Creek 1998)
- Occupations promote social inclusion. (Passmore 2003)
- The health and well-being of the individual are influenced by what he/she does. (Hasselkuss 2002)

In the early stages of Huntington's disease it may be appropriate to use a rehabilitative approach to interventions such as using a diary, calendar, wipe board to organise and prompt tasks. In the middle to late stages of the disease progression it is more appropriate to use a compensatory approach such as adapting the environment or changing routines.

Table 3: Highlights common impairments of people with Huntington's disease and their impact on activities and participation

**Table 3: Common impairments of people with Huntington's disease and their impact on activities and participation**

Physical Impairments	Problems with activity and participation
Chorea  Bradykinesia Dystonia	Difficulty maintaining posture and controlling limb movements impacting on all activities of daily living. Slowing of intentional movement. Contracting of specific muscle groups causing balance problems and increasing risk of falls.
Reduced manual dexterity, poor co-ordination, reduced grip	Difficulty with manual tasks such as eating, fastening buttons, writing, holding and operating a phone.
Dysarthria  Dysphagia	Loss of voice clarity and decreased intelligibility of speech. Difficulty swallowing food and drinks increasing the risk of choking and aspiration.
Fatigue	Reduced endurance affecting all tasks. Reduced ability to maintain posture and complete activities.
Cognitive Impairments	Problems with activity and participation
Bradyphrenia  Impaired executive function  Impaired attention  Reduced motivation  Depression Anxiety Dementia	Generalised slowing of thinking processes. Reduced problem solving and decision making skills. Reduced concentration and ability to maintain or switch attention. Apathy, reduced interest and drive leading to social isolation. Low mood, sadness, reduced motivation. Fear, avoidance of situations. Confusion and inability to maintain safety.
Behavioural impairments	Problems with activity and participation
Psychosis	Confusion and reduced concentration.
Challenging behaviour	Difficulty maintaining relationships. Inability to maintain safety.

## **x) Collaborative Working**

### **a) A multi-disciplinary team approach and multi-agency working**

When working with people with Huntington's disease and their families it is important to provide a cohesive and multi-disciplinary service. The disease affects many different aspects of the individual's abilities so they will need to be assessed by appropriately trained health care professionals for the various difficulties they experience. Aubeeluck (2009) states,

"The complex nature of HD makes it unlikely that any one professional will have all the skills needed to help any one individual. It is therefore of utmost importance that the service providers take a multi-disciplinary approach to HD in order to identify the best way to assist individual patients by taking into account their differing needs." (p360).

Whilst the person with Huntington's disease continues to live in their own home there can be a number of different agencies involved such as housing department, social work department, care agency workers, wheelchair services, employment services, day care centres. It can be difficult to keep track of the people involved and their contact details. For this reason, a care co-ordination approach works well where one person acts as a key worker and will liaise with the other professionals as necessary, advocate for the service user, and introduce new professionals to the service user at appropriate times. In some teams the key worker or care co-ordinator will be the occupational therapist.

### **b) Working with family/carers**

As an occupational therapist it is important to work with family, members, friends and informal carers as well as the professionals visiting the individual. The occupational therapist should be explaining the interventions that are being recommended to the family and carers so that they can prompt and encourage the person with Huntington's disease between visits. The occupational therapist can also provide information about the disease and its progression as appropriate and refer onto other members of the multi-disciplinary team if needed. Broaching the issue of environmental adaptations with service users and carers needs to be done sensitively and thoughtfully. Occupational therapists are aware of the progression of the disease, the adaptations which will probably be required and the length of time it takes to get these adaptations completed. Referral onto the local authorities for this work can appear urgent but needs to be managed with sensitivity. The individual and family may not be ready to accept the changes that the disease will cause and the changes which may be needed to their home. Sometimes a smaller piece of equipment will suffice in the short term but the occupational therapist may need to introduce the idea that, for example, a bath board will not work in the long term and that there are adaptations that can be undertaken when the family feel ready to discuss them.

### **c) Early and consistent intervention**

There are a number of advantages to early intervention with people who have Huntington's disease:

- Enables the therapist to build a therapeutic relationship with the service user and family at a point when the symptoms are relatively minor.
- Ideally the person will be reviewed regularly and then interventions can be increased when needed.
- Early intervention enables the use of rehabilitative strategies whilst the person retains the ability to learn and adapt to difficulties.
- Early intervention enables the therapist to know the service user well and to build trust which is important as the disease progresses and more professionals become involved and the decisions more difficult.
- If possible, establish and document any advanced decisions at an early stage such as medical care (e.g. PEG), management of finances (enduring power of attorney) whilst the individual has capacity to make these decisions. The Huntington's Disease Association's 'End Of Life' fact sheets has some useful information regarding end of life issues to consider (HDA 2009).

## **The Guidelines**

These guidelines have been written to cover all the stages of Huntington's disease. However, some challenges to occupational performance will be more prevalent at specific stages of the diseases progression.

## **1. Strategies for physical, cognitive and behavioural factors impacting on engagement, motivation and learning**

Huntington's disease typically sees deterioration in the 3 areas of cognition, physical function and mood/behaviour. This section describes some of the clinical signs and symptoms observed and how these impact on function. Suggestions of ways to overcome these problems are covered. Part 3 looks at facilitating engagement in more specific activities of daily living.

### **1.1 Physical factors**

Huntington's disease has a major impact on a person's physical function. The key changes that can occur are changes in gait and balance, involuntary movements (chorea), bradykinesia (slowness and delay in movement), reduced coordination, speech (dysarthria) and swallowing (dysphagia) impairment (Rosenblatt et al. 2000; Kirkwood et al. 2001).

An individual's physical presentation will change over the course of their illness. Clinical diagnosis is currently defined by the presence of motor symptoms as opposed to behavioural or psychiatric signs (Busse & Quinn 2009). One of the earliest physical signs may be chorea (involuntary movements (Quarell 1999). These are likely to be subtle at first and may not be particularly noticeable to the person with Huntington's disease. People may also begin to have problems with voluntary movement, e.g. bradykinesia. The motor rating test asks a person to perform a number of physical tasks aimed at trying to detect these changes. Although these subtleties may indicate that the disease has started, such physical issues may be less of a problem to the person, than the cognitive changes described later.

The presentation of Juvenile onset Huntington's disease differs from adult onset in that voluntary movement abnormalities, rigidity and spasticity may manifest itself early in the illness rather than later, and they may have more myoclonus (brief jerks of muscle groups). Thirty percent also have seizures (Quarrell & Brewer 2009; Rosenblatt et al 2000 in Busse & Quinn 2009).

As the disease progresses physical problems become more prominent. Chorea may become more obvious, with larger movements of the trunk, limbs and/or face observed. The bradykinesia will become worse, but may be masked by the presence of the chorea (Quarrell 1999). Dystonia may also become apparent. In some cases, people may experience chorea less and may become more rigid as the disease progresses. Falls may become more frequent as balance and gait become affected (Busse et al 2009). As the respiratory and bucolingual muscles are affected by the disease, speech will become slurred and more difficult to understand. The ability to perform a coordinated swallow also becomes more difficult (Heemskerk et al 2010). Incontinence and weight loss may also become an issue.

In the later stages of the illness the individual will become increasingly immobile and eventually will be no longer able to weight bear. Chorea may be severe or it may cease. At this stage, the person with huntington's disease has become totally dependent on others for their care. Choking becomes a major concern. People also eventually lose the ability to verbally communicate. Although they are generally still able to comprehend language, and retain an awareness of family and friends (HDA 2012).

**Guideline: Physical factors**

For occupational therapists aiming to assess and promote physical function for people with Huntington's disease, it is recommended that:

- The physical impairment of the person with Huntington's disease should be assessed. As several aspects of a person's physical function may be deteriorating, ascertaining where the physical impairment/s lies may enable task adaptation and facilitate engagement.
- Assess the impact of physical deterioration on activities of daily living (see Part 3).
- Use task analysis to break down the activity to assess whether the activity can be adapted to overcome physical and functional problems.
- Consider the environment and/or equipment used and whether this is facilitating or hindering a person's engagement and safety.

Where fatigue is an issue:

- Consider keeping interventions or activities short due to limited attention and fatigue.
- Consider the need to build in rest sessions into the daily routine.
- Consider which time of day the person has most energy.
- Activities are prioritised in conjunction with the person.
- Consider ways of preserving energy e.g. using a wheelchair on a longer trip.

Where communication is becoming difficult it is recommended that:

- Alternative methods of enabling communication should be established, as communication deteriorates.
- Be aware it can take time to 'tune in' to someone's speech. Consider working with a carer initially where possible.
- Where possible work in conjunction with a speech and language therapist to establish best strategies.
- Consider the person's ability to use a particular method and their likelihood of compliance. Low-tech solutions may be best e.g. yes/no cards, whiteboards and marker pens, alphabet charts. Consider the person's strengths such as writing ability, pointing, eye pointing.
- Consider establishing communication charts/folders with pictures or words of a person's known drink, food and activity preferences early on.

Where swallowing is becoming difficult it is recommended that:

- Therapists working with people with Huntington's disease should be aware of the risk of aspiration and/or choking, and should have awareness of the various signs that swallow has become affected e.g. gurgly voice, coughing and spluttering (see speech and language guidelines for more signs).
- Where a therapist becomes aware someone's swallow is affected, involvement of a speech and language therapist should be sought without delay. They can advise the person and their carer as to how to manage the condition.

## 1.2 Cognitive factors

There are a number of cognitive changes associated with the progression of Huntington's disease including: progressive impairment of attention, executive function, memory and psychomotor skill (Ho et al 2003).

### i. Executive function

Executive function is really an over-arching term for a host of functions which allow a person to plan and organise themselves over time, make complex and abstract judgements and to organise and control their memory processes.

In the early stages of Huntington's disease the pathological changes start in the dorsal caudate nucleus which forms part of the dorsolateral prefrontal cortex and from there gradually spreads throughout the frontostriatal system (Hedreen and Folstein 1995). Imaging studies have shown that impairments in executive function relate to this frontostriatal neuropathology. Executive dysfunction can cause a range of symptoms:

**Table 4: Commonly reported symptoms of frontal lobe dysfunction** (Stuss and Benson 1984, 1986)

Poor abstract thinking	Impulsivity	Confabulation
Poor planning	Euphoria	Poor temporal sequencing
Lack of insight	Apathy	Disinhibition
Variable motivation	Shallow affect	Aggression
Lack of concern	Perseveration	Restlessness
Can't inhibit responses	Lack of concern for social rules	Distractibility
Poor decision making		

Other symptoms associated with executive function that could be added to this list include initiation, multitasking difficulties, difficulties with spoken language, visual perception and motor control. There is not one single unifying explanation for these symptoms and therefore no single method of rehabilitation. Consequently, the clinician will need a range of techniques to draw upon according to each situation.

There are a number of specific interventions which can be used to help to overcome executive difficulties however, due to the progressive nature of Huntington's disease the approaches commonly used are environmental modification and compensatory strategies which involves changing the physical environment to simplify the executive loading of a task.

### ii. Attention and Psychomotor Skill

Attention has been sub divided into different areas such as focussed attention, divided attention and sustained attention. Although people with Huntington's disease may have difficulty with all of these areas the clinical term bradyphrenia, slowing of cognitive speed and capacity, probably best

describes the difficulties experienced by people with Huntington's disease. Using a connectionistic model, one mechanism that can explain this slowness is the 'detour effect', where a signal travelling through the network can no longer use the shortest route due a lack of or a-functional nodes (van Zomeren and Spikman 2008). In Huntington's disease this could be the damage to the sub-cortical circuits which decreases the speed of thinking. Haggard et al (2000) studied cognitive-motor interference; they observed a slowing in gait cycle in patients with brain damage who had to do cognitive tasks whilst walking. They concluded that dual task performance exceeded the available information processing capacity. This cognitive-motor interference can be observed in people with Huntington's disease who can lose their balance when walking and conversing at the same time as both cognitive and consciously controlled motor processes are competing for limited capacity.

### iii. Memory

People with Huntington's disease are prone to procedural memory deficits due to basal ganglia deterioration (Heindal et al 1989, Solveri et al 1997). Impairments in verbal episodic memory have been documented as well as impairments in learning and retrieval whilst storage is relatively spared. Solomon et al (2007) studied verbal episodic memory in people with the Huntington's gene but who had not yet been clinically diagnosed. Their findings indicate that verbal episodic memory is affected in pre diagnostic Huntington's disease.

In practice, many people with Huntington's disease appear to have good memories for past events but their working memory is possibly affected by their ability to understand and process information in the first instance and then to be able to initiate retrieval.

#### **Guideline: Cognitive factors**

For occupational therapists aiming to assess and promote the cognitive functioning for people with Huntington's disease, it is recommended that:

The person with Huntington's disease should be screened for cognitive impairment by use of functional and/or standardised assessment.

Consideration is given to the use of strategies to compensate for specific cognitive impairments highlighted in the assessment, such as:

- Use of lists, calendars, notebooks, wipe boards, sticky notes
- Establish a consistent daily routine
- Label drawers/cupboards around the home
- Use prompts to aid retrieval of information
- Offer specific choices e.g. rather than asking "what would you like to drink" ask, "would you like tea or coffee."
- Break down complex tasks into smaller sections.
- Write down tasks in sequential order to check understanding.
- Encourage completion of one step of task before moving onto next step.
- Use short sentences conveying one piece of information at a time and allow the person time to respond as a reply can be delayed. Use of closed questions, which require less effort to process, may help.
- Eliminate external stimuli wherever possible to enable focus of attention.

Occupational therapists educate the person with Huntington's disease and all family members or care givers regarding the cause and management of cognitive impairments.

### **1.3 Behavioural factors impacting on engagement, motivation and learning**

#### **i. Depression and Apathy**

Research into depression and apathy in Huntington's disease suggests that apathy is far more common than depression and that the two symptoms are not related (Naarding et al. 2009). Other studies have found that there is a strong correlation between apathy, executive dysfunction and decline in activities of daily living (Hamilton et al. 2003, Van Duijn et al 2007, Van Duikn et al. 2010). Depression is often associated with earlier stages of the disease and has been linked to greater self-awareness (Hoth et al. 2007). The prevalence of depressed mood in people with Huntington's disease has been shown to vary from 33% to 69% (Van Duijn et al. 2007). However, no relation between depressive symptoms and disease duration has been reported (Van Duijn et al 2007). There is an increased incidence of suicide reported in people in the early stages of Huntington's disease compared to the general population according to Morris and Scourfield (1996), they reviewed a number of studies from 1941-1985 and found the prevalence of depression in Huntington's disease varied from 9%-44%. There are a few hypotheses regarding the association between depression and Huntington's disease:

Coincidental- suggesting there is no difference in the rates of depression in people with Huntington's disease compared to those without but this does not agree with the research available.

Psychological- suggesting that the depression is a psychological response to the development of symptoms of Huntington's disease. However, researchers have found that in some cases depression has preceded any neurological symptoms (Folstein 1989).

Organic- this view suggests that depression is due to the neuropathological changes that occur in the brain and are the same as those causing the neurological changes.

Damage to the anterior cingulate-subcortical circuit has been linked to motivational disorders such as apathy and is likely to occur in Huntington's disease. According to Van Duijn et al (2007) apathy consistently appears to be positively related to disease progression and is strongly related to decline in everyday functional activities.

#### **ii. Deliberate Self-Harm**

There are also incidents where the individual is not depressed but due to cognitive changes thinks that suicide is the only solution to their situation therefore, it may be a behavioural response to particular circumstances. Other self-harming behaviours observed in people with Huntington's disease have been abuse of alcohol, overdosing on prescribed medication and refusal of food and drink.

#### **iii. Anxiety**

Some people with Huntington's disease report higher levels of anxiety than they experienced prior to onset of symptoms. The increase in anxiety would appear to be linked to the deterioration in

cognitive skills as people describe perseverative thoughts about specific things and some appear to lack patience or social skills because they have to say or do what they have thought of immediately before they lose concentration. Van Duijn et al (2007) reviewed the current research into the psychopathology in Huntington's disease gene carriers. They found that there were few studies that had explored the prevalence of general anxiety in people with Huntington's disease although in one study 61% of people at their first outpatient appointment reported experiencing anxiety. This is perhaps an area to be further researched in the future.

#### **iv. Psychosis**

Psychotic symptoms are found in approximately 10% of people diagnosed with Huntington's disease (Watt and Seller 1993, Pflanz et al 1991). Those people presenting with psychotic symptoms and no motor symptoms have been mis-diagnosed as having schizophrenia in the past (Dewhurst 1970 and Folstein 1989). Some authors have suggested that schizophrenia can be a precursor to Huntington's disease but Watt and Seller (1993) found that the onset of schizophrenia occurred around the time of onset of physical symptoms.

Morris and Scourfield (1996) report that paranoid psychosis may occur in Huntington's disease and Pflanz et al (1991) found that paranoid psychosis occurred in 5.9% of men and 7.7% of women with Huntington's disease. Van Duijn et al. (2007) suggests that the lower reports of psychosis nowadays is due to the better and earlier diagnosis of Huntington's disease and the shift in research from inpatient to outpatient populations.

#### **v. Aggression**

Carers of people with Huntington's disease report a high incidence of irritability and aggression, particularly in the mid stages of the illness. This behaviour is often linked to the cognitive changes occurring in the executive function of the brain as the disease progresses (Van Duijn et al 2007). The individual loses the ability to regulate their behaviour and may lack insight into the effects their behaviour has on family members or carers. It can be difficult for relatives to understand the changes in personality and to accept that the individual often cannot modify their responses. The increase in irritability and aggression has a direct impact on carer's stress levels and needs to be carefully monitored. The occupational therapist may work closely with the family to identify the triggers and escalators to the aggressive behaviour. It is then necessary to consider the options available to avoid or limit the triggers and to consider strategies to help the family to cope when the behaviour occurs. If a behavioural approach is not completely successful in managing these difficulties it is important for the occupational therapist to liaise with the doctor responsible for the person with Huntington's disease to review medical options. Sometimes a urine, ear or chest infection can have a marked effect on a person's behaviour, as can pain.

**Guideline: Behavioural factors**

For occupational therapists aiming to assess and promote the mental wellbeing for people with Huntington's disease, it is recommended that:

The person with Huntington's disease should be screened for emotional and neuropsychiatric impairment by use of communication, observation and liaison with family and/or caregivers.

Occupational therapists educate the person with Huntington's disease and all family and/or caregivers regarding the cause and management of emotional and neuropsychiatric impairments.

Occupational therapists refer person with Huntington's disease and the family members or caregivers to appropriate sources of support in relation to management of emotional and psychiatric impairments.

Occupational therapists promote the use of strategies to assist in the management of behavioural changes such as:

- Avoid confrontations
- Speak in a low soft voice
- Try to keep the environment calm and controlled
- Try to identify the triggers to irritability and avoid them
- Acknowledge the irritability as a symptom of frustration and respond diplomatically
- Stick to a regular daily schedule
- Encouraging the individual to focus attention on a new task (distraction) may be effective.

People in the middle stages of Huntington's disease may be less able to use psychodynamic or cognitive approaches to psychological interventions but may respond to a more behavioural approach.

## 2. Optimising activities

There is a lack of research and published information about the processes and techniques used by occupational therapists when treating people with Huntington's disease. Jain et al (2005) describe a practical framework of how an occupational therapist might intervene when treating people with progressive neurological conditions (see figure 1 below). This model could be used to organise interventions with someone with Huntington's disease and dynamically as the disease progresses.

Fig. 1. Framework for intervention

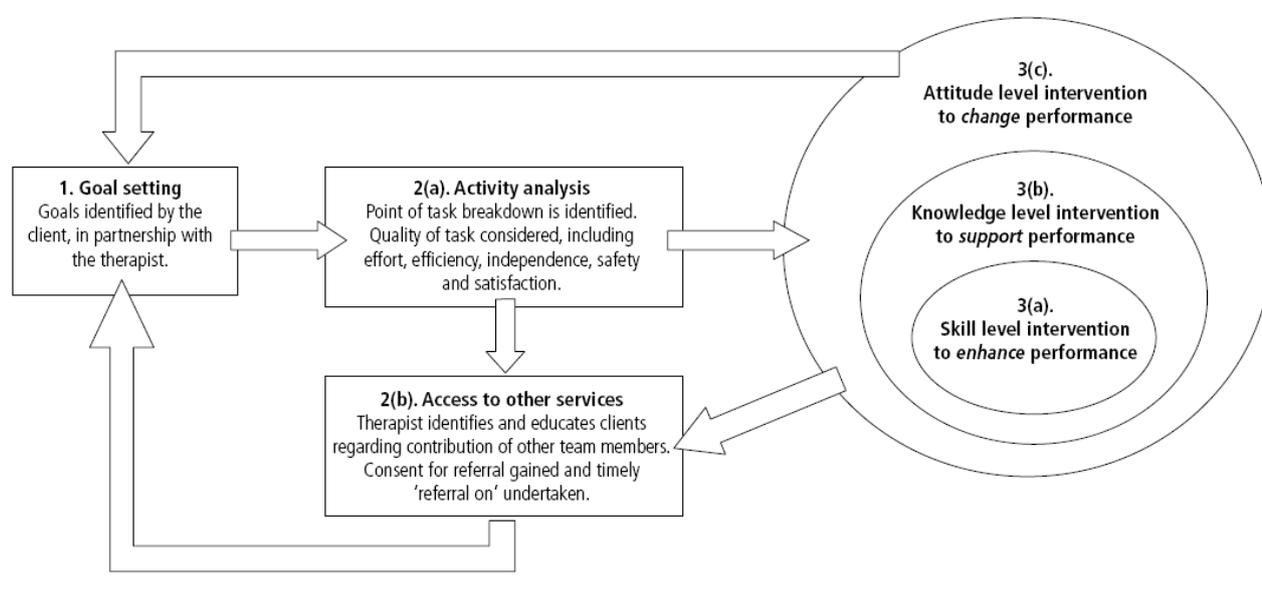


Diagram Jain et al (2005) inserted with permission from BJOT

The framework describes 3 separate, but interconnected steps to facilitate occupational performance.

### 1. Skill level intervention

This involves enhancing performance by improving people's skills. For example, teaching weight transference during sit to stand transfers or breaking a task down into small stages such as moving from lying to sitting by rolling onto one's side first with the goal of changing the pattern of movement used during bed transfers and hence reducing physical effort required.

### 2. Knowledge level intervention

This level of intervention aims to support performance by increasing knowledge about how to modify a task. Information about how to manage body level symptoms, access to equipment and resources can enable people to make informed choices.

### 3. Attitude level intervention

This level aims to change performance by modifying attitude and expectation. Here a task may be changed completely e.g. using a scooter for outdoor mobility, ordering shopping from the Internet, employing a cleaner.

#### **2.1 Optimising mobility and falls prevention**

Huntington's disease has a major impact on mobility. Involuntary movements, reduced coordination, prolonged muscle contractions and dystonia (abnormal posturing) can all result in impairment of balance and walking (Quinn et al 2009). A high proportion of people with Huntington's disease have recurrent falls leaving them at risk of injury (Busse et al. 2009).

Occupational therapists need to consider the increased risk of falls in a person's home and everyday activities. Assessment should include identifying potential hazards in the person's immediate environment. Building design may introduce risks, such as additional steps within the house or garden. Stairs are a major hazard for people with Huntington's disease as the illness progresses.

An additional stair rail may help some people, yet others tend not to use them. Lack of consistent hand grip can be an issue, reducing their usefulness. Consultation with physiotherapy colleagues may provide strategies to minimize risks on the stairs. Adaptations such as stair lifts need careful consideration because of the difficulties making controlled transfers at the top of the stairs. Through floor lifts take up lots of space and a person may bruise their limbs in a confined space if they have severe chorea. However there may be some situations where these are appropriate and would reduce risks. Reasons for suggesting such adaptations should be documented and their use reviewed regularly. Eventually it will be necessary to stop using the stairs and to adapt the ground floor to provide a sleeping and toilet/washing area.

Consideration of a person's usual activities and the way they conduct them may also highlight an increased falls risk. Task analysis may enable an activity to be adapted to reduce risk of exacerbating falls. For example, moving items out of low kitchen cupboards to reduce need for stooping. Also organising the environment to limit the amount of times a person has to change direction may reduce falls, as this commonly causes imbalance. Trying to do two things at once such as walk and hold a conversation may also diminish concentration and exacerbate falls.

Cleaning equipment that is heavy and cumbersome to use may exacerbate falls risk and alternatives should be considered (Haslam 2006). Equipment such as perching stools may stabilise a person during a task. However consideration needs to be given as to whether such equipment is then another hazard for someone to trip over, particularly if there is no one else to put it away.

As fatigue impacts on mobility, promoting the need to pace activity throughout the day and plan rest periods during the day, may assist. The need for mobility aids (such as a wheelchair) may need consideration. It is recommended that occupational therapists work closely with physiotherapy colleagues to manage mobility problems in the community. Any equipment to aid mobility will need full assessment of a person's physical and cognitive abilities and their behaviour.

Where falling is likely and unavoidable, a falls action plan, including a means of calling for assistance should be considered. How a person gets off the floor also needs to be addressed. Methods should be practiced and where assistance is required, suitable manual handling equipment and training be provided

**Guideline: Optimising mobility and falls prevention**

For occupational therapists aiming to promote mobility and reduce risk of falls in people with Huntington's disease it is recommended that:

A comprehensive MDT falls assessment should be completed and documented.

([www.cks.nhs.uk/falls\\_risk\\_assessment/management/scenario\\_falls\\_risk\\_assessment/interventions\\_in\\_falls\\_clinics](http://www.cks.nhs.uk/falls_risk_assessment/management/scenario_falls_risk_assessment/interventions_in_falls_clinics))

Assessment is undertaken to identify potential hazards in the person's immediate home environment.

Adapt the environment, where possible to reduce the risk of falls. Consideration should be given to potential hazards e.g. clutter, furniture, stairs, loose rugs, fireplaces, lighting.

Encourage the individual to adapt the way they conduct activities to reduce the risk of falls. For example,

- When carrying items suggest the items are placed in a pocket or a diagonal shoulder bag allowing the arms to remain free to stabilize on the walls or furniture

Identify and provide (where appropriate) a suitable means of raising the alarm in the event of a fall such as a mobile phone, careline or telecare system.

Educate individuals and carers on the need to concentrate when walking and limit distractions. For example, holding a conversation with a person who has Huntington's disease whilst they are walking can cause them to lose their balance.

Identify and practice a safe way of getting up from the floor in the event of a fall. This may be in conjunction with a lifting and handling specialist. Where moving and handling equipment is needed this should be provided without delay.

Review and monitor risk regularly as the person's condition changes.

## **2.2 Transfers**

Transfers can be a hazardous activity for people with Huntington's disease due to potential poor balance, coordination and chorea. Ability to transfer safely onto a chair, bed, toilet, and into a car, bath/shower, needs to be individually assessed.

Transfers can be heavy and uncontrolled. People can be observed to 'fall' in an uncontrolled manner, resulting in them being poorly positioned and causing potential risk of injury. Adapting the environment and/or equipment used, may go some way to reducing the hazards.

Where a bed is against a wall, clumsy transfers can result in risk of head injury on the nearby wall. Use of either a double bed to increase transfer space or moving the bed well away from the wall may reduce the risk. Where feasible, padding around the bed area may also assist in reducing injury.

Heavily transferring into a chair risks the chair moving or tipping up, and there is potential for the chair to break if it is not of a sturdy nature. Positioning furniture, e.g. placing a chair against a wall, may reduce tipping risk. However the chair must be high backed to avoid someone hitting his or her head against the wall behind. Sturdy furniture may reduce the likelihood of a chair moving or breaking. Where someone is using a specialist seating system, requesting brakes on all four castors may improve the chair's stability (Expert reference group 2010). To aid transfers off furniture, having beds/chairs set at the right height for the individual may go some way to facilitating independent transfers.

Heavy transfers onto toilets can cause the seat to loosen or break and the individual may be at risk of head injury on the cistern. Padding the area around the cistern may reduce risk of injury (Sulaiman 2007). Purchasing a toilet seat with sturdy bolts, set at right angles may reduce the frequency of the seat breaking.

Where possible, adapting the environment or equipment used to remove the need for challenging transfers should be considered. For example, replacing a bath with a level access shower, use of a rise recliner chair to facilitate sit to stand transfers. Where transfer aids e.g. grab rails and toilet frames are required, consider the need for them to be securely bolted or floor fixed to enhance their stability. Any equipment to facilitate transfers and ease caregiver burden needs individually assessing and a trial period undertaken.

For further information on transfers and physical management of Huntington's disease symptoms refer to Physiotherapy Guidelines (Quinn and Busse et al 2009).

**Guidelines: Transfers**

For occupational therapists aiming to promote and facilitate safer transfers of people with Huntington's disease it is recommended that:

A person's ability to transfer safely onto a chair, bed, toilet, and into a car, bath/shower, should be individually assessed.

Consideration should be given to adapting the environment and/or equipment used to reduce the hazards.

Consider the sturdiness of the furniture being used.

Beds/chairs should be set at the optimal height for the individual, to aid independent transfers.

The need for grab rails by the toilet and shower should be assessed and equipment installed without delay.

Where a toilet frame is deemed useful, it should to be floor fixed to enhance its stability.

Where possible, the environment should be adapted or equipment used to remove the need for challenging transfers. For example, replacing a bath with a level access shower. Where deemed suitable, use of a rise recliner chair may facilitate sit to stand transfers.

Any equipment (such as hoists, slide sheets, manual handling belts) to facilitate transfers and ease caregiver burden should be individually assessed and a trial period undertaken (see section 4.2).

All transfers should be regularly reviewed (the frequency may be determined by hospital policy), and amendments made as necessary.

### **2.3 Bed mobility and sleep safety**

As Huntington's disease progresses consideration needs to be given to a person's comfort, positioning and safety in bed and also the needs of carers in managing the person's needs.

Physical, cognitive and behavioural changes may result in some of the following difficulties:

- Difficulty getting in and out of bed.
- Difficulty moving in bed such as sitting up and changing posture.
- Slipping down in bed.
- Increased choreic movements may result in risk of limbs becoming trapped, friction sores developing.
- Falls from bed.
- Increased time spent in bed or reduced ability to change position in bed resulting in increased risk of pressure areas and contractions developing.
- Difficulty in obtaining/maintaining a position to reduce aspiration.

#### **Guidelines: Bed mobility and sleep safety**

**For occupational therapists aiming to promote bed mobility, sleep safety for people with Huntington's disease it is recommended that:**

- Individual risk assessment should be carried out on the suitability of the bed, mattress and any bedrails, in combination with the bed occupant. Document and review risk assessment regularly.
- In early stages, if a person is still transferring in and out of bed, and this is in an uncontrolled manner, consider padding around the bed area and removal of clutter. Having the bed at a suitable height to facilitate transfers is also essential.
- Consider teaching rolling over in bed and sitting up by breaking down the transfer into small stages. Such as moving from lying on back, on to side, up to sitting.
- Consider a person's cognitive as well as physical abilities such as ability to follow instructions, maintain grip or organise movements when assessing for equipment to aid transfers.
- When falls from bed are a risk, consider the use of a double bed or a bed which goes low to the floor.
- A falls mat may be considered following individual risk assessment. Consider the style of mat, could this increase the risk of an ambulant person tripping due to raised edges or a soft walking surface making the person unstable.

**Guidelines: Bed mobility and sleep safety**

- If bedrails are being considered they must be risk assessed on an individual basis. The person with Huntington's disease should be involved in the decision, where possible. It is very important to determine the views of the carers and nurses, as they will be the people using the equipment. If using bedrails, the risk assessment should be reviewed and update regularly. Refer to local policy on use of bedrails and National Patient Safety Agency (2007c) Safer Practice notice (17): Using bedrails safely and effectively and Medicines and Healthcare Products Regulatory Agency (2006) Device Bulletin 2006(6). Safe use of bedrails. Available at [www.mhra.gov.uk](http://www.mhra.gov.uk).

**For occupational therapists aiming to help maintain posture and reduce sliding down the bed:**

- Pillows and rolled towels can be used to offer support in maintaining posture.
- A profiling bed which has a 4 section profile action can reduce slipping by applying the knee break so that the knees are bent to reduce slipping down the bed. This must be considered when there is a need to elevate the head end of the bed.
- Consider the need for a profiling bed in the long term. This may facilitate transfers, aid positioning and be necessary for safe manual handling.
- The ability to profile a bed may aid respiration in the later stages and can minimise the risk of aspiration if a person is being PEG fed in bed or returns to bed for rest after a meal. For a person being PEG fed the minimum requirement is to sit at an angle of no less than 45 degrees (dietician guidelines) to reduce risk of aspiration.
- Consider the type of mattress, such as pressure relieving to suit the pressure relief requirements of the person and liaise with appropriate care professionals i.e. tissue viability nurse. Consider the future needs of the person. For example if a dynamic airflow mattress is required in the later stages, the bed would need to be compatible for this.

## **2.4 Postural Management**

Difficulty maintaining posture, especially when seated, may be problematic even at the point where somebody is mobilising independently. Sitting is a fatiguing activity, as the need to actively hold ones trunk erect requires good muscle tone and strength (Collins 2005). These aspects are frequently lacking in someone with such a disabling condition. Choreic movements may additionally impact on a person with Huntington's disease reducing their ability to maintain posture and balance.

Although each person with Huntington's disease will present differently there are some common problems encountered. Many people with Huntington's disease tend to slide down their chair resulting in sacral sitting, placing them at risk of developing pressure damage and reducing their functional ability. Leaning of the trunk to one side or rotation of the pelvis may be witnessed. In an effort to maintain an upright posture people may be seen to fix with their hands and elbows. As the disease progresses neck control may become problematic, which may result in flexion at the neck. Where someone is self-transferring, transfers are likely to be heavy and lacking in control. Injuries may occur if chairs are not well padded.

Ensuring seating is of appropriate dimensions is especially relevant for those with movement disorders (Aragon and Kings 2010). Failure to consider dimensions may exacerbate existing problems and will reduce success of any intervention (Collins 2005). A seat depth that is too long may contribute to sliding forwards. A seat that is too wide will allow the pelvis to move laterally or to rotate, which may contribute towards leaning. Ensuring a chair is measured and set up for the individual may go some way to alleviating problems seen.

Ensuring the person's feet are fully supported on the floor with footwear that provides grip may assist. Cushions can provide additional postural support and pressure relief. Therapists need to consider both and observe the person using the product to identify its suitability.

Some people may choose to lie on a sofa rather than sit. However it is important that people have the option of sitting more upright. A good posture is important when eating to reduce the risk of aspiration. The position of the head in relation to the shoulder girdle is the critical factor in facilitating swallowing, rather than an erect upright position of the body as a whole (Pope 2007). Periods of active sitting may also be necessary to delay further loss of muscle strength and tone, and will aid respiration (Pope 2007).

As the disease progresses it is likely that a more specialist seating system or individualised wheelchair, is required to enable people to achieve an upright central posture. Seating assessment should ideally be an interdisciplinary team process involving the patient. Where a tilt in space system is used be aware of the ability of people being able to keep their feet on footplates and risk of entrapment.

As people's mobility reduces and concerns about posture become apparent, a full review of posture and pressure care over a 24 hour period should be undertaken and reviewed as required (see part 4, end of life care).

**Guideline: Postural Management**

For occupational therapists aiming to promote improvements in posture and comfort in sitting for people with Huntington's disease it is recommended that:

An assessment of sitting ability and posture

Points to consider:

- Assess the dimensions of a person's current chair/seat to ascertain whether it is the correct size and offering adequate support.
- Assess whether the pelvis, trunk, neck, head and arms are adequately supported.
- Ensure the person's feet are fully supported. Footwear that provides grip may assist.
- Consider whether a cushion will provide additional postural support and whether pressure relief is required.
- Where people are unable to sit comfortably or safely consider the need for specialist lounge and/or wheelchair seating.
- Additional positioning devices may be considered to maintain position and/or safety. For example, head support, lateral trunk supports, positioning belts, a table or lap tray to provide upper body support.
- Least restrictive positioning methods and adaptations should be trialed first before deciding upon using a harness as a positioning aid. Where seating adaptations restrict freedom of movement (e.g. tilt in space, positioning harness) legal aspects regarding restraint must be respected. Consent should ideally be obtained from the person. Where this is not possible, the therapist should assess a person's capacity and where this is lacking they may need to act in a person's best interest. All such decisions need justifying and documenting as to the reasons for them. Consider whether a contoured backrest may also provide additional trunk support.
- Any seating system needs to be robust to withstand heavy transfers.
- Requesting brakes on all 4 casters may make transfers safer.
- Regular maintenance and checks of any seating/chair is necessary, as clips and screws can become loose.
- A modular seating or wheelchair system that can be adapted to increase its longevity can be useful where the individual's condition is changing frequently.
- Consider breathable materials to enhance comfort and wipe clean material due to continence and spillage issues.
- It is essential that a person's positioning and comfort should be regularly reviewed over time, as presentation will change. These options should be assessed in situ over time to assess whether they enhance posture and function.
- Training should be provided to either the person with Huntington's' disease and/or their carer on how to use any seating system/ adaptations, this should also be documented.

## **2.5 Eating and drinking**

People with Huntington's disease often report that they experience difficulties with eating and drinking. Due to the difficulties controlling movement the person with Huntington's disease often finds that they drop a lot of food during the course of a meal. This leads to frustration, embarrassment if eating in company, a reduced nutritional intake and fatigue. This can lead to a loss of interest in food and a withdrawal from social situations.

Many individuals with Huntington's disease have increased energy requirements and it is essential to provide adequate macro and micronutrients (Dietician Standards of Care Guidelines). A referral to a dietician should be considered so a baseline of nutritional assessment can be completed taking into account any increase of physical activity from chorea or a reduction of motor function in later stages of the disease.

Where the person with Huntington's disease has swallowing difficulties, such as coughing when eating, a gurgling voice or recurrent chest infections, a referral to a speech and language therapist should be made as the individual may benefit from a soft diet or thickened liquids. Some people with Huntington's disease choose to have a PEG (percutaneous endoscopic gastrostomy) as the effort and risks associated with swallowing become too great. Others may make an advanced decision not to have this intervention. This issue should be discussed with the multi-disciplinary team and the mental capacity of the person with Huntington's disease assessed to establish their ability to make the decision.

### **Guideline: Eating and drinking**

For occupational therapists aiming to promote eating and drinking abilities with people with Huntington's disease it is recommended that:

Where swallowing problems have been identified or the person is coughing on food and drink, a referral to a speech and language therapist should be made without delay.

Assess eating and drinking and provide equipment where appropriate as co-ordination of the fine movements coupled with involuntary movements and variable grip required to manage cutlery is problematic in Huntington's disease.

Strategies to assist with eating and drinking can also be suggested, such as:

- Use of spouted beakers and straws can help transportation of fluid to the back of the mouth and aid swallowing. However, if drinks are thickened the aperture size needs to be considered.
- Using a smaller spoon if the person with Huntington's disease puts too much food into his/her mouth.
- Appropriate visible snack foods in the kitchen or living room can prompt the person with Huntington's to eat if they have difficulty initiating the task.

**Guideline: Eating and drinking**

- Reduce the external stimuli when eating to reduce the risk of choking such as turning off the TV or not having a conversation.
- People become tired when eating and find it difficult to finish meals so smaller meals more frequently can help to insure the correct amount of nutrition. If carers take over feeding towards the end of a meal this can help to reduce fatigue.
- Those people who retain insight into the changes caused by Huntington's disease may become anxious about eating out because they tend to drop food or have difficulty chewing and swallowing. Some suggestions to assist with this are: ask to be seated at a table in a quieter area of the restaurant, choose a meal which will be easier to manage i.e. softer diet, or less effort to eat (penne rather than spaghetti), take adapted cutlery with you and a non-slip mat, arrange for everyone in the group to tuck napkins into their shirts/blouses.

## **2.6 Self Care**

People with Huntington's disease often report that their self-care routines become slower because of movement difficulties and the difficulty in concentrating and planning the tasks. Some people gradually stop initiating self-care routines and need prompting and support to manage. Discussions around the availability of care packages to assist with self-care should be undertaken with the individual and/or family and carers.

Dental/oral hygiene may become difficult for the person with Huntington's disease due to their movement difficulties and often a high calorie diet. They may need assistance and prompting to access a dentist/oral hygienist on a regular basis. Further guidance is available in the dental guidelines.

### **Guideline: Self care**

For occupational therapists aiming to promote self-care routines with people with Huntington's disease it is recommended that:

If self-care routines are lengthy or frustrating assistance should be provided in the morning and/or evening.

### **Toileting**

Assess for and provide handrails to assist transfers onto/off the toilet where appropriate without delay. Alternatively a toilet frame fixed to the floor.

Where appropriate, assess for and provide a padded backrest around the cistern if the person has strong extensor movements.

Where continence is a particular difficulty to the person with Huntington's and the caregiver a referral to the continence advisor /local nurse should be made without delay.

The choreic movements caused by Huntington's disease may result in the toilet seat breaking as the bolts wear over time. When purchasing a toilet seat, consider one with the sturdiest bolts or preferably one where the bolts insert at a right angle into the toilet seat. Be prepared to replace the toilet seat frequently.

Consider using a toilet that washes and dries.

### **Guideline: Self care**

#### **Bathing/showering**

Assess for and provide appropriate bathing equipment. The choice of equipment will depend on the person's symptoms, space in the bathroom and carer support. It is preferable to avoid wall mounted shower seats due to the risk of injury through hitting the head on the wall. Tilt in space shower chairs, shower cradles and easy access baths can be useful. In latter stages of the condition the person may need postural support in the bath. Training for carers in use of equipment should be regularly reviewed and documented.

#### **Dressing/grooming**

Assess and where appropriate suggest strategies to assist with dressing, such as:

- Loose fitting clothing with elastic waistbands are easier to manage, adapt to front fastening if appropriate.
- Zip tags on jackets can help.
- Under wired bras can be more uncomfortable for people in mid-late stage of Huntington's disease who are less active and sitting for extended periods of time.
- Be aware of weight loss and clothing falling down causing a trip hazard.
- Shoes should be supportive, lower heels for women are recommended and Velcro fastenings may be easier to manage.
- Clothes may need to be changed frequently due to increased soiling e.g. messy eating, drooling, and perspiration.
- If the person with Huntington's disease has difficulty initiating and organising the task consider laying the clothes out in order.
- Verbal prompts will be necessary as the condition progresses and even when another person is required to dress the person, simple commands can encourage the person with Huntington's disease to assist by lifting limbs.

If the person smokes, clothing that is slow to burn/melt is preferable. Fire retardant aprons and furniture throws are available if the risk of fire from smoking is high.

Assess and where appropriate suggest strategies to assist with grooming, such as:

- Using an electric shaver or beard trimmer is safer than a wet shave.
- Shorter hairstyles are easier to manage but for those people who wish to keep longer hair a 'tangle teaser' brush or spray in conditioner helps to keep hair manageable.
- Other aspects of grooming such as waxing, manicures and pedicures can often be arranged with local beauticians. It is important for carers to maintain these activities when the person can no longer do them themselves. It promotes social interaction and inclusion, promotes emotional well-being and is a sensory stimulating activity.

## **2.7 Domestic skills**

Performance of domestic tasks such as meal preparation, shopping and housework are affected during the mid to late stages of Huntington's disease. Cognitive changes relating to planning, organisation and decision-making impact on the ability to manage finances and running a household. However, these tasks can be a central role within the family for some people with Huntington's disease and important for them to maintain for continued self-esteem. Consideration should be given to modifying tasks for those people who wish to continue this role. For example, buying pre-prepared meals or pre-prepared vegetables, shopping on line or employing a cleaner.

Declining cognitive skills and a reluctance to request assistance can result in some people neglecting their property and failing to pay bills. If an individual with Huntington's disease does not have a friend or family member who calls and reports the problem the neglect continues to increase. The person may not be able to plan, organise or initiate tasks such as cleaning, checking the dates on food or organising household repairs. In extreme circumstances the individual can end up homeless. Other complications are dealing with bailiffs, repossession orders and finance companies. In recent years there has been much work in the UK around the implementation of the Mental Capacity Act 2005 and the safeguarding of vulnerable adults in healthcare trusts and local authorities. Gunstone (2003) studied the perceptions and experiences of mental health workers who assess and manage severe self-neglect. The findings suggest a number of 'grey areas' including a lack of definition of severe self-neglect and desensitising of mental health workers to neglect. Gunstone's participants defined self-neglect as including:

- Failure to maintain an adequate intake of food and drink.
- Failure to maintain personal care such as personal hygiene and wearing of appropriate clothing.
- Failure to maintain the home environment such as disposal of rubbish, keeping the home clean, health and safety issues such as poorly maintained appliances and fire risks.
- Failure to manage finances
- Failure to maintain social contact, in respect of accessing people to help them.
- Failure to comply with treatment, such as taking medication, attending outpatient appointments.
- Failure to protect themselves from abuse (sexual abuse, financial abuse, abuse of goods and property by others).

Where there is self-neglect, neglect of property or safeguarding issues the occupational therapist should work closely with the social worker, consultant, family and any other relevant professionals. The individual's mental capacity to decide how and when to care for themselves and their property should be assessed as well as their ability to carry out activities of daily living. If the individual lacks capacity a decision needs to be made which is the least restrictive and in their best interests (Mental Capacity Act 2005, Deprivation of Liberty Safeguards 2009).

**Guideline: Domestic skills**

For occupational therapists aiming to promote domestic skills and abilities with people with Huntington's disease, it is recommended that:

Small items of equipment should be assessed for and provided without delay. For example:

- Non-slip mats may ease jar opening and prevent plates slipping.
- Wire mesh baskets may help when draining vegetables.
- A trolley may be appropriate to assist with transferring objects.

Consideration should be given to breaking down domestic tasks into component actions to allow successful participation in some aspects of the activity.

Consideration should be given to introducing convenience foods and internet shopping where appropriate and support to plan shopping lists.

Carrying a meal or drink from the kitchen to the living room can be difficult to co-ordinate and increase the risk of falls. Where possible it can help to have a table and chair in the kitchen to eat and drink at. Some people manage to use a trolley. The metal type with larger wheels is preferable as it is sturdier and the larger wheels go over threshold strips more easily.

Consideration should be given to the use of a microwave rather than a conventional cooker. If using a conventional cooker, timers can be useful to remind the person to return to the task.

Consider reorganising food cupboards and freezer space and labelling drawers and cupboards can help people to find the items needed. Workspaces should be free of clutter and unnecessary items.

Smoke alarms should be fitted at strategic points within the home. The local fire service will usually assist with a fire safety assessment.

Occupational therapists should explore the need for assistance with or delegation of some or all of the household tasks such as ironing, housework, household maintenance, and management of paperwork/finances. This may involve a referral to local health and social care teams for a care package or involvement of the local authority appointee where finances are concerned.

## **2.8 Fatigue**

Little is written about the issue of fatigue in Huntington's disease. Studies have shown patients with Huntington's disease can have disrupted night-day activity patterns (Morton et al. 2005). Goodman et al (2010) in a study of sleep disturbance in Huntington's disease found various sleep-related difficulties were identified in a significantly greater proportion of Huntington's disease patients compared to control subjects. They conclude that disturbed sleep in Huntington's disease may contribute towards the deterioration of the patient's ability to perform activities of daily living and have a significantly deleterious effect on the quality of life of both patients and carers.

Whilst sleep disturbance is likely to impact on energy levels, engagement in activity may also result in physical fatigue. For example, those involved in physical activity (e.g. walking to the shops) can be observed to tire towards the end of the activity e.g. slowing of pace and increased stumbling may be evident. Also, when people have been engaged in a physical activity, some are observed to need to a period of rest after.

People with Huntington's disease can become physically fatigued which can affect their posture, particularly in the mid to late stages of the illness. During the course of the day, a person's ability to maintain their posture can decline, resulting in slipping in a chair. This may be mediated by a period of bed rest.

People with Huntington's disease can also become mentally fatigued. The concentration of people can be affected. In the mid-late stages people's ability to concentrate can be observed to dwindle, even when they engaged in a short activity. Skills of attention and processing information can also be affected by fatigue.

Some people in early stages of the disease will experience fatigue although they may appear to have no other symptoms.

However, whilst fatigue is observed to be an issue in Huntington's disease, it should be noted that the clinical picture is mixed. Whilst fatigue certainly affects some people, for other people it seems to be less of an issue.

**Guidelines: Fatigue**

For occupational therapists aiming to promote management of fatigue in people with Huntington's disease, it is recommended that:

A good sleep routine is promoted. Where sleep patterns are disturbed to a great extent discuss with the multidisciplinary team to review medication and management of sleep routine.

The impact of fatigue on performance in all activities of daily living should be assessed and/or measured.

Implementation of strategies to try and manage fatigue should be considered and trialled. For example, using a diary to record specific activities that increase fatigue or times of the day when fatigue is more of a problem.

Identify priorities and how best to use time and limited energy.

Identify energy conserving and labour saving strategies e.g. using a wheelchair/scooter for long distances, getting shopping delivered, delegating out tasks.

Planning activity to ensure energy is reserved for priority activities and allowing for rest periods after fatiguing activity.

Where mental fatigue is an issue, identifying types of tasks causing fatigue and investigating whether such tasks can be adapted/shortened or delegated to someone else.

Where postural fatigue is occurring, the length of sitting tolerance is identified and recommendations are made about periods of bed rest.

### 3. Supporting participation

The symptoms of Huntington's disease gradually develop over many years sometimes resulting in a marked impact on the person's quality of life. In addition to the physical, cognitive and behavioural changes caused by the condition the individual and family often have to deal with the environmental, psychosocial and financial challenges that accompany the progression of the condition. These may include:

- Accessibility of public places or operating equipment such as ATM or self-service checkout in the supermarkets.
- Being misunderstood by others who attribute their symptoms to being drunk or intoxicated by drugs.
- Feelings of embarrassment, stress and frustration at their changing abilities.
- Social withdrawal and isolation may occur as a response to social difficulties.
- Reduced means to fund the support required to maintain quality of life.

#### 3.1 Self Efficacy

Education of people with Huntington's disease and their family and/or carers to explain why they are experiencing difficulties with functional activities may be useful. Encouraging maintenance of physical activity, cognitive flexibility and engagement in social activities can prove beneficial in the long term.

Encouraging individuals and their families to adapt activities can promote positive coping methods. Maintaining work for as long as possible enables financial independence, role retention and self-worth.

#### **Guideline: Self efficacy**

For occupational therapists promoting self-efficacy and positive coping strategies with people with Huntington's disease it is recommended that they:

Assess participation restrictions affecting family, work, leisure and social roles.

Consider lifestyle planning and coping strategies to promote engagement in personally meaningful roles and activities.

Offer client-centred education, advice and information to promote participation and self-management.

Signpost to local services and peer support, including education, health promotion and self-help programmes such as the Huntington's Disease Association or carer support groups.

### **3.2 Roles and Relationships**

Changes in roles and relationships within families and couples are common when some or more members of the group have Huntington's disease. Williams et al (2009) studied the emotional experiences of family carers in Huntington's disease and found that carer's experienced the disintegration of their own lives, a loss of life as it had been and having to deal with the 'ever-present shadow' for those who had children/grandchildren.

Factors affecting roles and relationships may include:

- Partners feeling overburdened with responsibility for the household and care of the person with Huntington's disease.
- Depression, mood swings, agitation and behavioural changes in addition to communication problems can have an impact on family life.
- Loss of interest in sexual relationships or hyper sexuality can change the balance of a partnership.
- Loss of role as a worker/employee can create financial worries and loss of self-esteem for the person with Huntington's disease.
- There may be a lot of anxiety within families about the risk of inheriting the disease and concern about the possibility of passing the gene onto their children.
- Those family members not affected by the gene may experience survivor guilt and emotional numbness according to Tibben et al (1992).
- Carers may have no option but to reduce or stop working to care for a relative which can cause a major change in roles and financial difficulties.

Occupational therapists can gain insights into these sensitive issues when working closely with a family in their own home and may have a key role in supporting people in the maintenance of relationships.

#### **Guideline: Roles and relationships**

For occupational therapists aiming to support people with Huntington's disease in coping with the impact on their roles and relationships, it is recommended that they:

Promote maintenance of normal roles, daily routines and social habits by suggesting and practising task modification.

Act on health and well-being concerns of the whole family (including sexual relationships) without delay, and with consent by referring to any appropriate agencies such as Relate or Outsiders.

Occupational therapists should be aware that family members may not be aware of the genetic nature of Huntington's disease and that they may be at risk of inheriting the symptoms.

### 3.3 Work

Some people with Huntington's disease can continue to work for a number of years following the onset of symptoms. However, many individuals have difficulty in maintaining this role. McCabe et al. (2008) found that fifty-seven out of sixty-nine people with a degenerative neurological condition experienced a change from full-time work to unemployment. She reports that,

*"The main representation of this change of employment was that the patient simply could not continue to meet the demands of the position, although for some it was clear that they were either made redundant or forced to resign by their employers."(p604)*

McCabe et al (2008) also reports the impact on the individuals and their carers both financially and emotionally when work activities cease. Some people with Huntington's disease reported a relief to have finished work as they no longer had to cope with the stress but the majority of people reported the negative effects including loss of self-worth and identity leading to depression and anger.

There is some evidence to suggest that people with progressive neurological conditions value vocational rehab/support to manage performance of work tasks, assist in disclosure of condition to employers and to provide counselling and advocacy within the workplace (Sweetland et al. 2007).

Where appropriate occupational therapists can work with the employer and the individual with Huntington's disease to explain the impact of their symptoms and to plan any environmental changes required to enable the person to manage physically in the job and to consider the cognitive requirements of the job and how this can be managed if/when job tasks change.

Employers need to undertake risk assessments if the person's symptoms are impacting on health and safety issues in order to make any necessary adjustments. There are schemes such as Access to Work which can assist to support people within the workplace. Information on this scheme and other work issues is available at the department for work and pensions website [www.dwp.gov.uk](http://www.dwp.gov.uk).

Additional information available in Work Matters: Vocational Navigation for Occupational Therapy Staff published by the College of Occupational therapists.

**Guideline: Work**

For occupational therapists aiming to address work-related issues with people with Huntington's disease, it is recommended that:

The occupational therapist provides support and information about work retention. This should be offered to enable diagnosis disclosure at the right time for the individual.

The occupational therapist should adopt the pro-work advocacy role, providing a link between the individual, the workplace and government services such as Access to work or the Disability Employment Advisor.

If practical problems are being experienced at work a work assessment should be undertaken to establish key physical and cognitive difficulties.

Reasonable workplace adjustments are made to enable the person with Huntington's disease to physically and cognitively undertake their job.

Guidance about the employer's role and responsibility under the Disability Discrimination Act (2004) should be given.

If the person with Huntington's disease decides to stop work the emotional, practical and financial impacts need to be explored and discussions held with the employers, human resources and/or trade unions, to establish the most favourable terms and timing.

**3.4 Social Recreational and Leisure activities**

For the disabled person who is no longer able to work, leisure plays an even more important part in daily living. Leisure activities may provide opportunities to fill time, be creative, maintain or increase social contacts and can be a substitute for work (Foster 1996).

The physical and mental benefits of social interaction and physical activities have been documented (NICE 2008). In a recent study, Trembath et al (2010) found age of onset of the illness was associated with a more passive lifestyle irrespective of a patients CAG repeat length. Those with more passive lifestyles experienced onset of the illness 4.6 years earlier than those people who were more active. They conclude being engaged in active pastimes may slow onset of the disease.

Studies show Huntington's disease to have a detrimental effect of people's recreational activities. McCabe et al (2008) found the majority of participants in their study to indicate that the illness had a major negative impact on recreational activities. Changes included reduced amount of social and physical activity, restrictions to hobbies, difficulties going on holiday, difficulty playing and in some cases, watching sport. These changes affected not only those with the illness, but also their

carer. Helder et al (2001) assessed the impact of Huntington's disease on health related quality of life. Findings indicated severest impairment to be in the psychosocial dimensions of the sickness impact profile (including work, recreation & pastimes, home management). They recommend directing attention to the psychosocial well-being of people with Huntington's disease in clinical practice.

Changes to lifestyle brought on by the illness, may affect ability to engage in recreational activity. McCabe et al (2008) found opportunity for a social life was affected by a reduction or cessation of work. Reduced finances may also impact on ability to afford particular leisure opportunities. Aspects of the illness itself will also affect people's ability to engage in leisure activities. People with Huntington's disease often appear apathetic. However this may be the result of reduced ability to plan and initiate activity (Quarrell 1999).

Cognitive changes, such as reduced concentration and memory may make previously enjoyed pastimes more difficult e.g. reading, watching films. Motor changes (e.g. reduced balance, impaired grip, chorea, rigidity,) and fatigue, may also impair ability to engage in various leisure pursuits. Research in this field has demonstrated that those with disabilities withdraw from all but the most passive occupations (Blacker et al 2008).

Thorough assessment is required to ascertain what recreational/social activities are important to someone with Huntington's disease, the reasons why these pursuits are affected, and how. Involvement of a family member/carer may be helpful where people are unable to identify what is important to them or where they are unable to communicate this.

Where a person wishes to continue with a pursuit/pastime, occupational therapists should consider whether the activity could be adapted to continue successful and safe participation. Adaptation may be adaptive equipment, assistive technology or simply conducting a task in a different way. Choice of environment may have an impact on an individual's ability to participate in an activity e.g. a quiet environment to minimize distraction may assist someone with attention difficulties to engage (Blacker et al 2008). Where someone no longer wishes to continue with a pursuit or it is no longer practicable, occupational therapists may be able to work with the person to assist them in identifying or trying other recreational options.

The Huntington's disease association organizes nationwide support groups and fundraising events in the UK. This may be a useful source of support and information for people.

In the later stages of the illness when many activities may become too cognitively or physically demanding, sensory stimulation (e.g. music, massage) can provide a means of engaging people in pleasurable activity. Leng et al. (2003) found improvements in mood and behaviour in people with Huntington's disease, in response to multisensory environments. Facilitating people's ability to access the community/outdoors (e.g. through wheelchair provision) is also important in the later stages to maximise quality of life.

**Guideline: Social Recreational and Leisure activities**

For Occupational therapists aiming to promote social, recreational and leisure activities with people with Huntington's disease it is recommended that;

A person with Huntington's disease should be encouraged to use some of their time and energy to participate in social and recreational aspects of daily life, to promote mental and physical well-being.

Social, recreational and leisure priorities should be investigated as part of a holistic occupational therapy assessment.

Involvement of a carer/relative may be useful, where a person lacks insight, is apathetic or where communication difficulties exist. Interest checklists can be useful.

Occupational therapists should also consider the impact of caring for someone with Huntington's disease on a carer's social and recreational activities and whether this can be mediated.

Occupational therapists should consider whether an activity could be adapted to continue successful and safe participation. Consider whether adaptive equipment, assistive technology or conducting a task in a different way facilitates engagement.

Occupational therapists should consider whether the environment may impact on an individual's ability to participate in an activity e.g. a quiet environment to minimize distraction may assist someone with attention difficulties to engage (Blacker et al 2008).

Where someone no longer wishes to continue with a pursuit or it is no longer practicable, occupational therapists may be able to work with the person to assist them in identifying or trying other recreational options.

Access to local services for people with Huntington's disease and their carers should be signposted. The Huntington's Disease Association holds information on local branches and groups.

In the later stages of the illness, sensory stimulation (e.g. music, massage) may provide a means of engaging people in pleasurable activity and should be considered.

Wheelchair provision (by referral to wheelchair services) enables people at all stages of the disease to access the outdoors which is important to maximise quality of life.

### **3.5 Driving**

People who are symptomatic and have a positive diagnosis of Huntington's disease must notify the DVLA in Swansea in order to comply with the law. Failure to notify the DVLA can lead to penalty points, disqualification and a fine. Failure to notify the DVLA can also invalidate the vehicle's insurance. The individual is also responsible for notifying the car insurance company of the diagnosis. This should not affect the premiums but if not informed, the insurance company may not pay in the event of an accident <http://www.hda.org.uk/download/fact-sheets/HD-Driving.pdf>

The DVLA medical assessment unit liaises with the individual's GP and consultant and may make one of the following decisions:

- Restrict the person's driving license for 1, 2 or 3 years enabling a regular medical review to take place.
- Request a re-test.
- Revoke the license.

#### **Guidelines: Driving**

For occupational therapists who identify that a person with Huntington's disease is a motor vehicle driver, it is recommended that:

Information about the law and driving should be provided. Information available from <http://www.hda.org.uk/download/fact-sheets/HD-Driving.pdf>

If the individual's fitness to drive is questionable this must be raised with the individual and/or their family immediately. All discussions should be documented and the relevant GP or consultant informed.

Consider a referral to a driving assessment centre for a comprehensive assessment.

Occupational therapists should provide information about disabled parking schemes, usually available from local councils.

### **3.6 Community living skills and outdoor mobility**

Enabling people with Huntington's disease to participate in activities outside the home has a positive effect on physical and mental well-being. However, McCabe et al (2008) report that 19% of patients and 88% of carers reported that their illness had adversely affected their recreational activities. Some of the reasons that people find it difficult to access community and recreational activities are lack of suitable and affordable transport and lack of support needed to participate in activity due to severity of symptoms.

#### **Guidelines:** Community living skills and outdoor mobility

For occupational therapists aiming to promote opportunities to participate in community life and outdoor mobility for people with Huntington's disease, it is recommended that they:

Assess the individual's road safety skills and develop management strategies.

Encourage use of pedestrian crossings wherever possible as difficulty judging speed of traffic, processing information and initiating movement can increase risks of injury.

Introduce shop mobility schemes and supervise practice as required.

Provide information about the RADAR toilet scheme.

Make timely referral to local wheelchair services.

Provide written or verbal reminders of items to take when going out such as wallet/purse, keys, phone, travel pass where necessary.

Ensure that external grab rails and path rails are in situ as required between the home and public highway.

Suggest that the individual optimises their mobility by avoiding the shops at busy times.

With the person's consent, it may be useful to liaise with staff at the recreational facility (leisure centre, pub, club, café, etc.) to explain the support the individual needs or who to contact should help need to be provided.

Assess ability to transfer safely in/out of transport. Consider whether the individual can maintain a safe posture in seat and whether the driver is at risk.

Consider accessing community transport schemes.

### **3.7 Advanced Care Planning**

Advance care planning is the process of planning ahead when it is known that an individual has a deteriorating condition. There are two aspects of advance care planning that need to be considered.

#### **Advance statements**

This is a statement of the individual's wishes, preferences and plans for the future. It is not legally binding but can be essential in the process of adapting to the situation and increasing the sense of control for the future.

#### **Advance decisions**

This is a legally binding document about medical treatment the individual does not wish to happen. It requires the individual to have an assessment of mental capacity [www.legislation.gov.uk](http://www.legislation.gov.uk), [www.justice.gov.uk/guidance/...the.../mental-capacity-act/index.htm](http://www.justice.gov.uk/guidance/...the.../mental-capacity-act/index.htm)

Decisions which may have to be made by the person with Huntington's disease and their family as the condition progresses are:

- Whether to have the surgical procedure (PEG) to insert a tube into the stomach when the person has dysphagia or is losing weight.
- Whether to request a 'do not resuscitate (DNR)' policy when and if the person is admitted to hospital.

The National Gold Standards Framework in the UK provides information and tools to assist in advanced care planning and the end of life care pathway.

[http://www.goldstandardsframework.org.uk/About\\_GSF](http://www.goldstandardsframework.org.uk/About_GSF)

### **3.8 Lasting Power of Attorney**

Some people with Huntington's disease have a Lasting Power of Attorney (LPA) which is a legal document registered with the office of the public guardian and allows the person to nominate one or more people to make decisions on their behalf when they no longer have capacity to do so.

There are two different types of Lasting Power of Attorney:

- Health and welfare Lasting Power of Attorney – a health and welfare Lasting Power of Attorney allows the person with Huntington's disease to choose one or more people to make decisions for things such as medical treatment. A health and welfare Lasting Power of Attorney can only be used if the individual lacks the ability to make decisions for themselves.
- Property and financial affairs Lasting Power of Attorney - A property and financial affairs Lasting Power of Attorney lets the person with Huntington's disease choose one or more people to make property and financial affairs decisions. This could include decisions about paying bills or selling their home. An attorney to look after property and financial affairs can be appointed at any time. A condition can be included that means the attorney can only make decisions when the individual loses the ability to do so themselves. ([www.directgov.uk](http://www.directgov.uk))

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Occupational therapists need to know if the individual has a power of attorney nominated as they need to be consulted when treatment is being offered when the individual no longer has capacity to consent. The person nominated to act on the individual's behalf should also be involved in decisions about alternative living arrangements.

## **4. End of Life Care**

There are a number of treatments and medications that can be used to help control some of the symptoms associated with Huntington's disease, however at present there is no way to stop or reverse the course of this condition. As Huntington's disease is a neurological condition of long duration it may have a lengthy palliative phase. (Travers 2007).

The Promoting Excellence in End-of-Life Care Huntington's Disease Workgroup defines the initiation of palliative care as the point at which independent living is no longer possible (Klager et al 2008). While many people affected by Huntington's disease will be cared for at home, a large number will require institutional care as the disease progresses and they become increasingly more debilitated. This care can take place in a wide range of health and social care settings such as care homes, psychiatric units, acute hospitals, hospice care and care of the elderly units.

Palliative care is 'an approach that improves the quality of life of patients and their families facing life-threatening illness, through the prevention, assessment and treatment of pain and other physical, psychosocial and spiritual problems' (WHO 2011). There is evidence to suggest that clients and families affected by Huntington's disease may be in need of palliative care early on in the disease (Skilbeck and Payne, 2005 in Travers 2007). As people become cognitively impaired as the illness progresses, their ability to be fully involved with decisions about their care can become compromised. Delaying the introduction of a palliative care approach could affect decisions and wishes of the person, particularly around end-of-life care.

Acknowledging the patient's past history and preferences, keeping the family aware of new therapeutic options, and providing anticipatory guidance can facilitate each transition during this progressive disease (Moskowitz 2001). The National end of life care programme (2011) suggests that the following care principles should be included in end of life care:

- Care is client-centred and integrated
- Treat individuals with dignity and respect
- Identify and respect people's preferences
- Provide care after death.

(National end of life care programme, 2011)

Although there is little evidence demonstrating the efficacy of occupational therapy at the palliative or end of life stage of for people with Huntington's disease, occupational therapists have an important role to play in this context and may be involved throughout the steps of the end of life care pathway.

- Step 1. Discussions as the end of life approaches
- Step 2. Assessment, care planning and review
- Step 3. Co-ordination of care
- Step 4. Delivery of high quality services in different settings
- Step 5. Care in the last days of life

- Step 6. Care after death.

The End-of-Life Care Huntington's Disease Working group has delineated several priority areas for patient care in Huntington's disease: autonomy; dignity; meaningful social interaction; communication; comfort; safety and order; spirituality; enjoyment, entertainment and well-being; nutrition; and functional competence (Klager et al 2008).

Occupational therapists can focus on improving quality of life by promoting and identifying opportunities for enjoyment of free time (e.g. sensory activities such as music and outings), and through facilitating access to the outdoors e.g. wheelchair provision. Risks associated with increasing physical dependence and immobility will also need to be assessed and managed such as ensuring appropriate positioning and pressure care (National end of life care programme, 2011).

#### **4.1 24-hour postural management**

In the later stages, posture management needs considering on a 24-hour basis. Research suggests that regular changes to position over a 24-hour period are necessary for maintaining muscle length and preventing contracture formation in people with progressive neurological conditions (Pope 1992, 1997, Goldsmith 2000 in Aragon 2010). Pressure risk should also be measured and managed over a 24-hour period (NICE 2005).

Whilst assessment of sitting posture is important to ensure poor posture isn't contributing to pressure damage, a person's posture whilst lying also needs to be considered. Where people are unable to change bed position themselves damage can occur. Many asymmetries in body posture may originate in lying and then are compounded in sitting (Pope 1997).

**Guideline: Posture management**

For occupational therapists aiming to address end of life or palliative care with people with Huntington's disease it is recommended that:

A comprehensive review of posture and positioning over a 24-hour period should be undertaken.

A full posture assessment in both seating and lying is undertaken.

Where a person cannot change position themselves in bed the need for regular change of position by carers and support in lying must be considered.

A suitable 24-hour positioning regime should be implemented, including advice on managing positioning in: bed, lounge seating, wheelchair and car/vehicle.

Suitability of a person's lounge seating should be regularly reviewed and adapted or changed to meet altering needs.

Regular review of a person's wheelchair suitability should be undertaken and where necessary referral made to wheelchair services without delay.

Assessment of whether altering wheelchair or lounge chair set up (e.g. amount of tilt in space) can assist in maintaining good posture over time should be undertaken and advice given as necessary.

A full MDT review of seating must include the assessment of risk of pressure sores using a valid and reliable rating scale (NICE 2005). Results must be recorded with a clear review date.

Pressure care products which cater for a person's risk level e.g. cushions, should be considered and pressure care mattresses should be provided without delay.

#### **4.2 Manual handling and minimizing risk**

The manual handling needs of someone in the later stages of the illness are considerable. As people become increasingly immobile, provision of a suitable hoist and slings is likely to be necessary. The need for more manual handling manoeuvres e.g. bed positioning, will also become evident.

Careful consideration needs to be given to the equipment used, its suitability for the person with Huntington's disease and the environment it will be used in. The number of caregivers required to carry out a task also needs ascertaining.

##### **Guideline: Manual handling**

For occupational therapists aiming to address manual handling issues in the later stages of Huntington's disease it is recommended that:

A review of all transfers and manual handling scenarios throughout a 24 hour period should be undertaken.

Any equipment required should be sought without delay.

Any equipment deemed useful should be initially trialled in situ and training provided for caregivers on its use.

The number of staff required to transfer a person safely and position them satisfactorily needs to be established.

All recommendations should be documented and reviewed regularly in conjunction with the carer(s).

Where skin integrity is an issue, the fabric type of slings should be considered.

Due to the likely lack of head and neck control in the later stages, slings that provide maximum support around the head and neck, should be considered.

As involuntary movements or resistance may make transfers hazardous, padding around the hoist boom should be considered.

Due to the complexities involved in late stage Huntington's disease occupational therapists should consider working with other relevant professionals (e.g. physiotherapists, and/or manual handling trainers) to discuss manual handling issues.

### **4.3 Alternative living arrangements**

As occupational therapists we aim to enable people to continue living in their own homes for as long as possible, if this is their choice. However, as Huntington's disease progresses there comes a time when additional support is needed within the home and for many people a specialist nursing home placement is required. Proactive care planning, family support and discussion around the type of care available can help to avoid a breakdown of the home situation and the need for emergency hospitalisation.

Living arrangements need to be reviewed regularly as caregiver stress can increase immensely as the Huntington's disease progresses. It is important to inform the family and caregivers about other sources of care available in their area and how this is funded. Care can be provided but often takes several weeks to get the appropriate assessments and paperwork completed before funding is approved therefore, as occupational therapists it is important to explain this process so that appropriate referrals can be made before crisis point is reached by the family.

In addition to referring on to appropriate agencies once additional care is needed the occupational therapist may have a role in the following areas:

- Liaising with the social worker or continuing healthcare advisor about the individual's needs and how their symptoms impact on their function and the family. Families can find it difficult to explain the extent of the cognitive difficulties or the severity of the behavioural or psychiatric changes in their relative.
- Once a care agency is identified the occupational therapist may need to work with the agency carers to explain how Huntington's disease affects the individual and the most effective ways to provide care for that person.
- To provide information about specialist nursing homes in the area which provide 24 hour care for people with Huntington's disease.  
(National End of life care Programme, 2010)

Where possible it is important to advocate choice for individuals and families regarding care agencies and nursing home provision.

**Guideline: Alternative living arrangements**

For occupational therapists aiming to address alternative living arrangements with people with Huntington's disease, it is recommended that:

Manual handling risks and burden of care must be reviewed regularly.

Concerns about health and well-being of the family (including children) should be acted on without delay and with consent of those involved.

Make a referral without delay to local authority where safeguarding is an issue for children who are at risk of neglect or abuse.

The team should meet with the person with Huntington's disease and their family to discuss concerns about risk, health and well-being and to raise options for external support, respite care, alternative accommodation or placement.

Information should be presented in a sensitive and understandable way, with written information provided.

Emotional and practical support should be offered to the individual and family members to help them to make decisions concerning changes in living arrangements.

Assessment of mental capacity of the individual with Huntington's disease should be undertaken as required. For those who lack the capacity to make decisions about their accommodation the team and family members need to work together to make a decision which is in the individual's best interests.

Reassessment of the person with a 24-hour care routine must be undertaken once they are living elsewhere and equipment provided without delay.

# **Appendix 1**

## **Literature Review**

Review of available literature relative to Occupation therapy in Huntington's disease.

### **Databases searched:**

MEDLINE 1950.... to February Week 1 2010  
EMBASE 1974 ...to January Week 3 2010  
CINAHL 1981 .....to January Week 3 2010  
AMED 1929..... to January Week 3 2010  
PsychINFO 1806.....to January Week 3 2010  
NEED searched February Week 1 2010  
Cochrane Library searched February Week 1 2010  
Internet searched February Week 1 2010

### **Search Strategy:**

Articles published in English: Therapy in human Huntington's disease.

Subjects only: Subjects to be of 18 years of age + with a confirmed diagnosis of Huntington's disease.

### **Search terms included:**

Huntington's disease, AND  
Occupational therapy / occupation / independence / rehabilitation / activities of daily living / adaptations / home adaptation / equipment / leisure / work / employment / feeding / posture / seating / wheelchairs / splinting / sensory / multi-sensory / manual handling.

A systematic literature search was performed electronically using the databases mentioned above. On occasion references of potentially relevant articles were also obtained from articles reference lists and followed up. This was done to seek and evaluate evidence of occupational therapy used with patients with Huntington's disease, and of any specific occupational therapy interventions which form part of the practice of clinicians from the standards of Care Group. All relevant publications were identified and categorised to evidence statements (see table 1). Due to the limited scope of the literature found, observational studies were included in the review process. Qualitative studies were also included and reviewed separately. A summary of the literature can be found in appendix A. Due to the apparent lack of scientific evidence recommendations were also formulated based upon clinical experience and expert consensus from within the European Huntington's Disease Network (EDHN) Standards of Care Occupational Therapist group.

**Table 1 Summary of Evidence for Occupational Therapy in HD.**

1++	<u>High quality meta-analyses, systematic reviews of RCTs or RCT's with a very low risk of bias.</u>
1+	<u>Well conducted meta-analyses, systematic reviews or RCTs with a low risk of bias.</u>
1-	<u>Meta-analyses, systematic reviews, or RCTs with a high risk of bias.</u> Leng T, Woodward M J, Stokes M, Swan A, Wareing L, Baker R. <a href="#">Effects of Multisensory Stimulation in people with Huntington's Disease: a randomized controlled pilot study.</a> Clinical Rehabilitation (2003): 17 30-41
2++	<u>High quality systematic review of case control or cohort studies</u>  Bilney B, Morris M E, Perry A. <a href="#">Effectiveness of Physiotherapy, Occupational Therapy and speech Pathology for People with Huntington's Disease: A systematic Review.</a> Neurorehabilitation and Neural Repair 17 (1)(2003)
2+	<u>Well conducted case control or cohort studies with a low risk of confounding or bias and a high probability that the relationship is causal.</u>
2 -	<u>Case control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal.</u>  Zinzi P., Salmaso D., De Grandis R., Graziani G., Maceroni S., Bentivoglio A., Zappata P., Frontali M., Jacopini G. <a href="#">Effects of an intensive rehabilitation programme on patients with Huntington's disease: a pilot study</a> Clinical Rehabilitation 21(7):603-13, 2007 Jul.

3	<p><a href="#"><u>Non-analytic studies e.g. case reports, case series.</u></a></p> <p>Mason J, Andrews K, Wilson E. <a href="#"><u>Late Stage Huntington's Disease: Effect of Treating Specific Disabilities</u></a> British Journal of Occupational therapy (1991) 54 (1)</p>
4	<p><a href="#"><u>Expert Opinion</u></a></p> <p>Blacker D, Broadhurst L, Teixeira L. <a href="#"><u>The Role of Occupational therapy in leisure adaptation with complex neurological disability: A Discussion using two case study examples.</u></a> Neurorehabilitation 23 (2008) 313-319</p> <p>Fenech A, Baker M. <a href="#"><u>Casual Leisure and the Sensory Diet: A concept for improving the quality of life in neuropalliative conditions.</u></a> NeuroRehabilitation 23 (2008) 369-376</p> <p>Aubeeluck A <a href="#"><u>Management and Therapies for HD-related dementia</u></a> British journal of Healthcare management (2009), Vol 15 No 4.</p> <p>Higgins D S. <a href="#"><u>Huntington's Disease</u></a> Current Treatment Options in Neurology (2006), 8 (236-244).</p> <p>Abeeluck A <a href="#"><u>A holistic and multidisciplinary approach to Huntington's disease management</u></a> International Journal of Therapy and Rehabilitation 2009 16:7360-361</p>

	<p><b>Myers C</b> <a href="#"><u>The many faces of Huntington's disease. Part II: Occupational therapy's role in improving quality of life</u></a> <b>Advance for Occupational Therapy Practitioners 2007 4 32-34</b></p>
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As there is no mechanism for incorporating qualitative studies in the evidence base, and is an area of development of the SIGN Guidelines, those considered relevant are listed separately below.

McCabe M P, Roberts C, Firth L.

[Work and recreational changes among people with neurological illness and their caregivers.](#)

Disability and Rehabilitation (2008): 30 (8) 600-610

Zinzi P., Salmaso D., De Grandis R., Graziani G., Maceroni S., Bentivoglio A., Zappata P., Frontali M., Jacopini G.

[Effects of an intensive rehabilitation programme on patients with Huntington's disease: a pilot study](#)

Clinical Rehabilitation 21(7):603-13, 2007 Jul. (2)

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Leng T, Woodward M J, Stokes M, Swan A, Wearing L, Baker R.

[Effects of Multisensory Stimulation in people with Huntington's Disease: a randomized controlled pilot study.](#)

Clinical Rehabilitation (2003): 17 30-41

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Pilot Study  Randomised Controlled 2 group design	Specialist residential unit for people with mid- late stage HD.	HD Group N=12  N =6 Treatment group (MSE) group – 1 pt dropped out  N=6 Control group (relaxation) 1 pt dropped out  Mid – late stages  All functioned at Shoulson & Fahn's stage 5.  2 changes in medication during study.	4 week base line period for all pts  4 week intervention period 2 x sessions a week 30 minutes per session Individual treatment sessions in same room off the ward.  4 week follow up period  Treatment group Multisensory environment (MSE) used visual, tactile, auditory and olfactory stimulation  Control group Relaxation used listening to music and being read to.	Ratings made at 2 wk intervals using;  REHAB (Hall & Baker)  BMD (behaviour & mood disturbance scale)  BMD also done pre, during & post sessions.  Physiological assessment; BP, HR, RR. (Done once on 3 days per week during intervention period)	No Significant difference found between treatment group & control on main outcome measures.  MSE group shared the within session effects- Significant changes in stimulation & mood levels. This immediate effect not sustained.  - As sessions increase a cumulative effect seen in stimulation level and mood.  Findings suggest a leisure but not therapeutic effect.

Steultjens E NJ, Dekker J, Bouter L, Leemrijse C, Van den Ende C HM.

[Evidence of the efficacy of occupational therapy in different conditions: an overview of systematic reviews](#)

Clinical rehabilitation (2005) 19: 247-254

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Overview of systematic reviews	N/A	14 systematic reviews included.  1 was a review of studies of OT with people with HD	Review conducted in Pubmed (1966-Oct 2004) & Cochrane library (October 2004)  Reviews included were those that utilised a systematic search for evidence with regard to OT for for specific pt groups.	Descriptive analyses of studies found.	No conclusion could be stated on whether OT was effective in improving outcomes for HD, due to small no. of studies with small sample sizes & poor quality methodology.  Evidence exists for OT's ability to maintain & improve function for stroke, elderly & RA patients.

Mason J, Andrews K, Wilson E.

Late Stage Huntington's Disease: Effect of Treating Specific Disabilities

British Journal of Occupational therapy (1991) 54 (1)

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Multiple baseline single-case experimental design	Inpatient specialist HD unit	N= 4 with HD  Late stages Dependent for daily care but considered potential to assist with some ADL.	8 month study  4 Activities of Daily Living (ADL) randomly allocated 2 treatment ADL 2 ADL acted as a control.  3 phases –  1) baseline observation period 8 wks 2) 16 weeks ADL ax & intervention – at 2 weekly intervals 3) 8 wk- Follow up Ax of chosen ADL.	Components of task identified & each rated according to performance on a 9 point scale – dependent – independent.  Participants rated for each component part. Scores combined to give overall score for task.	No consistent change in treated ADL conditions. 1 patients abilities fluctuated  However, only 1 patients abilities deteriorated.  Possible generalised effect of OT in preventing deterioration.

Bilney B, Morris M E, Perry A.

**Effectiveness of Physiotherapy, Occupational Therapy and speech Pathology for People with Huntington's Disease: A systematic Review.**  
**Neurorehabilitation and Neural Repair 17 (1)(2003)**

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Systematic review of literature examining outcomes of OT, speech & physiotherapy for people with HD	N/A	Patients had confirmed diagnosis of HD 18 yrs +	<p>English published electronic data bases searched; CINAHL 1982-2001, Embase 1984- 2001, Medline 1966 -2001, Cochrane 2001, Allied and Complementary Medicine 1985-2001and PEDro database for physiotherapy trials 2001.</p> <p>Observational studies which examined treatment and interventions by physiotherapists, occupational therapists and speech pathologists</p> <p>Interventions were included if they were provided for impairments of movement, cognition or emotional status and aimed to improve the performance of activities or participation in society.</p> <p>Methodological quality assessed.</p> <p>Expert opinion identified but not included in data analysis</p>	Articles found were graded for study type using a hierarchy of levels of evidence	<p>Search failed to find any studies ranked as level 1, 2 or 3 evidence.</p> <p>3 articles found for OT in HD. Insufficient evidence to draw treatment recommendations</p> <p>2 observational single case design;</p> <p>1) Mason et al – No gain in function found, but OT may of prevented further deterioration.</p> <p>2) Di Scipio &amp; Hannesson – characterised by lack of objective valid reliable outcome measures.</p>

Fenech A, Baker M.

Casual Leisure and the Sensory Diet: A concept for improving the quality of life in neuropalliative conditions.

NeuroRehabilitation 23 (2008) 369-376

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Descriptive case study	Specialist in-patients HD unit.	1 with HD  Severe cognitive dysfunction	Individualised sensory diet programmes encompassing all 7 senses & based on detailed sensory profile described.  -Vestibular stimulation: coach trips, car rides, outings – seaside, pub, tourist attractions -Visual stimulation: bubble tube, fibre optic light strands, films on TV, drama, dancing & live entertainment -Olfactory stimulation: aromatherapy massages, aromas & oils -Tactile stimulation: table games & creative activities; art pottery indoor gardening. Sun on skin -Gustatory stimulation: Spicy and oriental food, tea & hot chocolate -Auditory stimulation: discussion group & current affairs, audio books, concerts & musical events -Proprioceptive stimulation: aromatherapy massage, shiatsu, & Aquability.	Observation & self report	Patient reported feeling satisfied with leisure choices.  Using sensory diet as a guide to plan casual leisure opportunities may provide way to engage patients with severe cognitive function & broaden sensory leisure experience.  Further research is required.

Blacker D, Broadhurst L, Teixeira L.

[The Role of Occupational therapy in leisure adaptation with complex neurological disability: A Discussion using two case study examples.](#)

Neurorehabilitation 23 (2008) 313-319

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
2 case studies	Inpatient specialist HD unit	1 with HD 1 with ABI  Patient with HD Age = late 40's Mid stages  Clinical features affecting function; Generalized chorea affecting upper/lower limbs, fatigue, dysarthria	Client centred creative leisure activity.  OT intervention;  Stabilisation of posture. Adaptation of environment & task – limited session time & Low tech equipment.	Observation & length of time spent in the activity	Adapting activity appeared to minimise fatigue, optimise participation time & promoted functional use of upper limbs.  Enabling participation contributed to participants sense of achievement, opportunity to maintain social & communication skills.

Zinzi P., Salmaso D., De Grandis R., Graziani G., Maceroni S., Bentivoglio A., Zappata P., Frontali M., Jacopini G.

[Effects of an intensive rehabilitation programme on patients with Huntington's disease: a pilot study](#)

Clinical Rehabilitation 21(7):603-13, 2007 Jul.

Design	Setting	Participants	Intervention/ Methods	Outcome Measures	Findings and Conclusions
<p>Pilot Study</p> <p>Intervention</p> <p>Within subjects design</p>	<p>Inpatient rehabilitation programme</p>	<p>HD Group (HD): N=40 (M=17)</p> <p>Early- mid stages as per UHDRS TFC (all symptomatic and received diagnosis of HD prior to enrolment)</p> <p>Age= 52. 0 s.d.= 3.3</p> <p>Absence of severe psychiatric disease based on DSM IV</p> <p>MMSE&gt;20</p> <p>CAG repeat 40-54</p>	<p>Individualised treatment programme included respiratory exercises, speech therapy, physio, OT and cognitive rehabilitation.</p> <p>3 weeks of intensive treatment repeated up to 3 times a year. 8 hrs a day for 5 days.</p> <p>Individual &amp; group sessions</p> <p>OT aimed at learning new strategies to perform tasks by stimulating memory, planning strategies &amp; prolonging concentration. Involved in swallowing, walking, standing up in safe way, promoting safety in the environment &amp; autonomy in eating, personal hygiene &amp; dressing.</p>	<p>- Zung Depression Scale</p> <p>- Mini-Mental State Examination</p> <p>- Barthel Index</p> <p>Completed before each admission period.</p> <p>Tinetti Scale</p> <p>Physical Performance Test (PPT)</p> <p>Completed on admission and on discharge.</p>	<p>Each 3 weeks period of treatment resulted in highly significant (P&lt;0.001) improvements of motor performance and functional performance.</p> <p>No carry over effect from one admission to the next was apparent, but at the same time no motor decline &amp; no functional decline was detected over 2 years - indicating that patients maintained a constant level of function, motor and cognitive performance.</p>

Busse, M A, Rosser A E.

**Mobility and falls in people with Huntington's Disease**

Journal of Neurology, Neurosurgery & psychiatry (2009) 80: 88-90

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Observational Study	Hospital clinic	<p>N = 24 HD</p> <p>Mean age = 56.6</p> <p>Score of 4 on the UHDRS motor diagnostic confidence rating.</p>	<p>Patients assessed using Berg Balance Scale (BBS) &amp; Timed Up and Go (TUG).</p> <p>Walking confidence (using ABC-UK) &amp; walking speed assessed.</p> <p>Walking activity recorded using 'step watch step activity monitor' worn for 7 consecutive 24hr periods.</p> <p>Questionnaire used to collect data on falls &amp; stumbles over past 12months.</p>	<p>Berg Balance test (BBS)</p> <p>Timed up &amp; go test (TUG)</p> <p>Unified UHDRS Functional Assessment scale(FAS)</p> <p>Independence scale</p> <p>Total functional capacity (TFC)</p> <p>Data on stumbles &amp; falls.</p>	<p>TUG &amp; BBS predicted probability of falling &amp; may be suitable for use in HD.</p> <p>Confirms people with HD fall regularly.</p> <p>Falls management therefore needs to be priority.</p> <p>As falls likely to be multi factorial in origin varying factors e.g. home modification / medication need further investigation.</p>

Aubeeluck A

Management and Therapies for HD-related dementia

British journal of Healthcare management (2009), Vol 15 No 4.

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Opinion					<p>A multidisciplinary approach optimises effectiveness of care for the patient.</p> <p>OT can help with progressive physical disabilities &amp; home adjustments.</p>

McCabe M P, Roberts C, Firth L.

Work and recreational changes among people with neurological illness and their caregivers.

Disability and Rehabilitation (2008): 30 (8) 600-610

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Individual interviews  Qualitative	Participants own home or work or via telephone.	<p>People with neurological illness and their caregivers.</p> <p>24 with Huntington's Disease.</p> <p>Age range = 43-68 yrs. Time since diagnosis = 1-9 years.</p> <p>In addition 28 professionals including OT's who worked with these populations were included.</p>	<p>Individual interviews. Majority conducted face to face. Completed by 1 researcher.</p> <p>Standard questions asked about -changes that occurred in work &amp; recreation as a result of illness - patient &amp; carer reactions to this.</p> <p>Further follow up questions developed during study.</p>	<p>Transcripts analysed by qualitative statistical analyses program NVivo 2.0. Coded based on themes.</p> <p>Themes identified by 3 separate researchers &amp; agreed.</p> <p>Triangulation used to agree on final themes reported.</p>	<p>6 major themes emerged under which findings were;</p> <ol style="list-style-type: none"> <li>1) Impact on patients work - 57/69 became unemployed.</li> <li>2) on carers work – 55 % became part time, 15 unemployed.</li> <li>3) Patient feelings about work changes – 55% indicated negative feelings (financial strain, isolation, depression).</li> <li>4) carers feelings – 60% indicated negative feelings.</li> <li>5) Impact of work changes on patients social life – 28 % negative changes (lack of finance)</li> <li>6) on carers social life – 30% report negative changes (isolation)</li> </ol> <p>The study findings highlight the importance of professionals in facilitating workplace maintenance &amp; addressing carers needs.</p>

Zinzi P., Salmaso D., Frontali M., Jacopini G.

Patients and caregivers' perspectives: assessing an intensive rehabilitation programme and outcomes in Huntington's disease

Journal of Public Health (2009), 17 (331-3380)

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Questionnaire Post treatment.	N/A	HD Group N=40  Mild – moderate HD.  All had completed at least 1 course of intensive inpatient rehabilitation in previous 3 years.  Av age = 49 yrs.	Structured Questionnaire to gain subjective evaluation of rehab program.  Questionnaire included multiple choice, dychotomic & open questions about aspects of treatment  Including; - Evaluation of duration of improvement - Attitude towards program - Rating rehab experience on 0-10 scale. - Evaluation of organisation aspects		37 returned. 93% response rate.  100% of respondents reported overall positive effect of treatment.  71% estimated benefits lasted 1-3months.  Perceived improvements in motor & functional performance match quantative outcome measured in 2007 study. Recall bias – possibility. Cannot establish individual part of OT

Higgins D S.

Huntington's Disease

Current Treatment Options in Neurology (2006), 8 (236-244).

Design	Setting	Participants	Intervention/Method	Outcome measures	Findings/Conclusions
Opinion	N/A				<p>Potential of rehabilitation is often overlooked in symptom management.</p> <p>Acknowledgement that wheelchair assessment, workplace modification, driving assessments, home evaluation for safety &amp; functional needs play a role.</p>

Myers C

The many faces of Huntington's disease. Part II: Occupational therapy's role in improving quality of life

Advance for Occupational Therapy Practitioners 2007 4 32-34

Design	Findings/Conclusions
Opinion	<p>Occupational Therapy (OT) interventions often overlap with physical therapy.</p> <p>OT specifically address</p> <ul style="list-style-type: none"> <li>- Activities of Daily Living (ADL) performance</li> <li>- assess for adaptive equipment needs</li> <li>- provide compensatory strategies for cognitive loss</li> <li>- recommend home adaptations</li> <li>- educate on home safety</li> <li>- carry out care giver training</li> <li>- may address swallowing difficulties if have expertise in dysphagia</li> <li>- positioning/seating systems</li> <li>- contracture management</li> <li>- environmental adaptations</li> </ul> <p>Stage of HD progression will change emphasis of OT intervention</p> <p>OT in Early Stages Myers describes:</p> <ul style="list-style-type: none"> <li>- a fitness program including strengthening core muscles and major muscle groups, higher balance activities, education on changes in sensation &amp; perception, aerobic and coordination exercises</li> <li>- strategies for cognitive changes; memory, planning &amp; concentration strategies</li> </ul> <p>OT in Middle stages:</p> <ul style="list-style-type: none"> <li>- relaxation techniques to reduce muscle stiffness and continue with exercises</li> <li>- compensate for spatial deficits : 3 part sequence when sitting down 1 touch chair before sitting down 2 turn around 3 sit down</li> <li>- rollator walker for walking</li> <li>- fatigue management; rest periods, use of wheelchair</li> <li>- stability in ADL i.e. eating and adaptive equipment</li> </ul> <p>OT in Late Stages:</p> <ul style="list-style-type: none"> <li>- Safety regarding: falls, impaired self awareness, uncontrolled movement, skin breakdown, impulsivity</li> <li>- Provide opportunities for walking with assistance, frequent position changes, range of motion exercise and splinting, frequent meals, bed/chair alarms to alert carer the potential for getting up and safe sleeping environment, chairs for positioning and offer tilt and recline.</li> </ul>

Abeeluck A

A holistic and multidisciplinary approach to Huntington's disease management

International Journal of Therapy and Rehabilitation

2009 16:7360-361

Design	Findings/Conclusions
Opinion	<p>HD can be managed by focussing on management and therapy in maintaining function and reducing the effects of the disease.</p> <p>Quality of life for the patient can be improved with 'the implementation of an individual care-plan that is based on a full assessment of the patient in the context of their disease state, their family and social environment'.</p> <p>Input from 'Occupational therapy and physiotherapy to help with progressive physical disabilities and home adjustments.</p> <p>Patients can be missed by professionals as the illness 'crosses many service boundaries'.</p> <p>A Multidisciplinary approach is needed to manage HD; medical, nursing, social and allied health care.</p> <p>A clear understanding of these services and 'communication between professionals and with the patient and their family is key'.</p>

## References

Aragon A and Kings J (2010) *Occupational therapy for people with Parkinson's. Best practice guidelines*. College of Occupational Therapy. London.

Aubeeluck A (2009) A holistic and multidisciplinary approach to Huntington's disease management, *International Journal of Therapy and Rehabilitation*, 16 (7), 360.

Baron K, Kielhofner G, Lyenger A, Goldhammer V, and Woenski J (2006) *The Occupational Self Assessment (OSA) A User's Manual*, University of Illinois At Chicago The Model of Human Occupation Clearinghouse Department of Occupational Therapy.

Baum CM (1995) The contribution of occupation to function in person's with Alzheimer's disease, *Journal of Occupational Science*, Australia, 2(2): 59-67.

Blacker D, Broadhurst L, Teixeira (2008) The role of Occupational Therapy in leisure adaptation with complex neurological disability: A discussion using two case study examples. *Neurorehabilitation* 23, 313-319.

Braveman B, Robson M, Velozo C, Kielhofner G, Fisher G, Forsyth K, and Kerschbaum J. (1998) *Worker Role Interview (WRI) A User's Manual*, University of Illinois At Chicago The Model of Human Occupation Clearinghouse Department of Occupational Therapy.

Buchan T. (2002) The impact of language and culture when administrating the Assessment of Motor and Process Skills: A case study, *British journal of Occupational Therapy*, 65, 371-373

Busse M, Wiles C M, and Rosser, A E (2009) Mobility and falls in people with Huntington's Disease. *Journal of Neurology and Neurosurgery & Psychiatry* 2009; 80 88-90

Chard C, Tram C, Liu L, and Mulholland S, (2006) *Verbal cueing and environment modifications; strategies to improve engagement in occupations in persons with Alzheimer's Disease*. Poster presentation WFOT 2006.

Chung J (2004) Activity, Participation and well-being of people with dementia in long-term care settings. *OTJR: Occupation, Participation and Health*, 24 (1)

Collins, F, (2005) The Ibis XP tilt in space powered wheelchair: A review. *International Journal of Therapy and Rehabilitation*, 12, (11), 510-514

Creek J (ed.) (1998) *Occupational Therapy: new perspectives*. Whurr, London.

Darragh A, Sample P, Fisher AG. (1998) Environment effect on functional task performance in adults with acquired brain injuries: Use of the Assessment of Motor and Process Skills. *Archives Physical Medical Rehabilitation* 79, (4), 418-423

Dewhurst K (1970) Personality disorder in Huntington's disease. *Psychiatrica clinica* 3:221-229. In Harper PS (ed) (1996) *Major problems in neurology: Huntington's disease*, second edition, Saunders, London.

Folstein S (1989) *Huntington's disease: A disorder of families*. Baltimore, MA: John Hopkins Press.

Foster M (1996) Life Skills. Chapter 8 in Turner A, Foster M and Johnson S (1996) *Occupational therapy and physical dysfunction. Principles, Skills and Practice* 4th Ed. London. Churchill Livingstone.

Gern A (1993) *Validity of the Worker Role Interview*. Master Thesis, University of Illinois at Chicago

Goldsmith S (2000) Postural care at night within a community setting. *Physiotherapy*, 86 (10), 528-534 in Aragon A & Kings J (2010) *Occupational Therapy for people with Parkinson's Best Practice Guidelines*. London. College of Occupational therapists.

Goodman AO, Morton AJ, and Barker RA (2010) Identifying sleep disturbances in Huntington's disease using a simple disease-focused questionnaire. *PLoS currents*, 2010, vol./is. 2/(RRN1189), 2157-3999 (2010)

Gunstone S (2003) Risk assessment and management of patients whom self-neglect: a 'grey area' for mental health workers, *Journal of psychiatric and Mental Health Nursing*, 10, 287-296.

Haggard P, Cockburn J, cock J, Fordham C, and Wade D (2000) Interference between gait and cognitive tasks in a rehabilitating neurological population. *Journal of Neurology, Neurosurgery and Psychiatry*, 69, 479-86.

Hamilton JM, Salmon DP, Cory-Bloom J, Gamst A, Paulsen JS Jerkins S, Jacobson MW and Peavy G (2003) *Journal of Neurology Neurosurgery and Psychiatry*, 74, 120-122.

Haslam R and Stubbs D (2006) *Understanding and preventing falls*. Taylor & Francis . CRC Press.

Hasselkuss B (2002) *The meaning of everyday occupation*. Slack, New Jersey.

Hedreen JC and Folstein SE (1995) Early loss of neostriatal striosome neurons in Huntington's disease. *Journal of Neuropathol Exp Neurol* 54: 105-120

Heindel WC, Salmon DP, Shults CW, Walicke PA, Butters N (1989) Neuropsychological evidence for multiple implicit memory systems: a comparison of Alzheimer's, Huntington's and Parkinson's disease patients, *Journal of Neuroscience*, 9, 582-7.

Helder D I , Kaptein A, Van Kempen, GMJ Van Houwelingen JC, & Roos RAC (2001) Impact of Huntington's disease on quality of life. *Movement Disorders*, 16(2), 325-330.

Ho AK, Robbins AOG, and Barker RA (2003) Huntington's disease patients have selective problems with insight, *Movement Disorders*, 21(3), 385-389.

Hoth KF, Paulsen JS, Moser DJ, Tranel D, Clark LA and Bechara A (2007) Patients with Huntington's disease have impaired awareness of cognitive, emotional and functional abilities, *Journal of Clinical and Experimental Neuropsychology*, 29 (4), 365-376.

Huntington's Disease Association web site. [www.hda.org.uk](http://www.hda.org.uk) Accessed 23 Jan 2011.

Jain S, Kings J and Playford ED (2005) Occupational Therapy for people with progressive neurological disorders: unpacking the black box, *British Journal of Occupational Therapy*, 68(3), 125-130.

Kielhofner G, Malinson T, Crawford C, Nowak M, Rigby M, Henry A, and Walens D (2004) Occupational performance history interview II version 2.1

Kirkwood SC, Su JL, Conneally M and Fouroud T (2001) Progression of symptoms in the early and middle stages of Huntington's disease. *Archives of Neurology*, 58, 273-278.

Klager J, Duckett A, Sandler S, and Moskowitz C. (2008) Huntington's disease: a caring approach to the end of life. *Care Management Journals*, 9(2), 75-81.

Law M, Baptiste S, Carswell A, McColl MA, Polatajko HJ, Pollock N (1994) *Canadian Occupational Performance Measure*. CAOT, Toronto

Leng T R, Woodward M J, and Stokes M J (2003) Effects of Multisensory Stimulation in people with Huntington's disease: a randomized controlled pilot study. *Clinical Rehabilitation* 17: 30 -41.

Marion C, Kagel MA, Norman A and Leopold DO (1992) Dysphagia in Huntington's Disease: A 16-Year Retrospective. *Dysphagia*, 7, 106-114.

McCabe M P, Roberts C, and Firth L (2008) Work and recreational changes among people with neurological illness and their caregivers. *Disability and Rehabilitation*. 30(8) 600-610.

Medicines and Healthcare Products Regulatory Agency (2006) Device Bulletin 2006(6). Safe use of bedrails. Available at: [www.mhra.gov.uk](http://www.mhra.gov.uk) (accessed 25.10.2010)

Moore-Corner R, Kielhofner G, and Olson L. (1998) *Work Environment Impact Scale (WEIS) A User's Manual*, University of Illinois At Chicago The Model of Human Occupation Clearinghouse Department of Occupational Therapy.

Morton AJ, Wood NI, Hastings MH, Hurelbrink C, Barker RA, and Maywood ES (2005) Disintegration of the sleep-wake cycle and circadian timing in Huntington's disease. *Journal of Neuroscience*, 25(1), 157-63,

Moskowitz CB, and Marder K (2001) Palliative care for people with late-stage Huntington's disease. *Neurologic Clinics*, 19(4)849-865.

Murgod UA, Saleem Q, Anand A, Brahmachari SK, Jain S, Muthane UB (2001) A clinical study of patients with genetically confirmed Huntington's disease, *Indian Journal of Neuroscience*; 190, 73-78

Naarding P, Janzing JGE, Eling P, van der Werf S and Kremer B (2009) Apathy is not depression in Huntington's disease, *Journal of Neuropsychiatry and Clinical Neuroscience*, 21 (3), 266-270.

National End of Life Care Programme <http://www.endoflifecareforadults.nhs.uk>

National Gold Standards Framework  
<http://www.goldstandardsframework.org.uk>

National Institute for Health and Clinical Excellence (2005) *Pressure ulcers: The management of pressure ulcers in primary and secondary care*. London: National Institute for Health and Clinical Excellence, available at <http://www.nice.org.uk/CG035>

National Institute for Health and Clinical Excellence (2008) Occupational therapy interventions and physical activity interventions to promote the mental wellbeing of older people in primary care and residential care. London: National Institute for Health and Clinical Excellence, available at: [www.nice.org.uk/nicemedia/pdf/PH16Guidance.pdf](http://www.nice.org.uk/nicemedia/pdf/PH16Guidance.pdf)

Passmore A (2003) The occupation of leisure: three typologies and their influence on mental health in adolescence. *OTJR: Occupation, Participation and Health*, 23(2): 76-83.

Perrin T (1998) Lifted into a world of rhythm & melody. *Journal of Dementia Care*, 6 (1): 22-25

Pflanz S, Besson JOA, Ebmeier KP and Simpson S (1991) The clinical manifestation of mental disorder in Huntington's disease: a retrospective case record study of disease progression. *Acta Psychiatrica Scandinavica* 83, 53-60, in Harper PS (ed) (1996) *Major problems in neurology: Huntington's disease*, second edition, Saunders, London.

Pope PM (1992) Management of the physical condition in people with chronic and severe neurological pathologies. *Physiotherapy* 78(12) 869-903 in Aragon A and Kings J (2010) *Occupational Therapy for people with Parkinson's Best Practice Guidelines*. London. College of Occupational therapists.

Pope PM (1997) Management of the physical condition in people with chronic and severe neurological conditions living in the community. *Physiotherapy* 83(3), 116-122, in Aragon A and Kings J (2010) *Occupational Therapy for people with Parkinson's Best Practice Guidelines*. London. College of Occupational therapists.

Pope PM (2007) *Severe and complex Neurological Disability management of the physical condition*. Butterworth Heinemann Elsevier. Philadelphia.

Rosenblatt A, Ranen N, Nance M and Paulsen J (2000) *A Physician's guide to the management of Huntington's disease*. New York, Huntington's disease Society of America.

Quarrell O, (1999) *Huntington's Disease – The facts*. Oxford University Press. Oxford.

Quinn L, Busse M, (2009) *Physiotherapy Guidance Document*. European Huntington's Disease Network, Germany.

Shanahan D, Evans A (2009) An audit of bedrail use and implications for practice. *British journal of Nursing*, 18(4), 232-237

Shoulson I and Fahn S (1979) Huntington's disease: clinical care and evaluation. *Neurology* 29(1), 1-3.

Solveri P, Brown RG, Jahanashi M, Caraceni T, and Marsden CD (1997) Learning manual pursuit tracking skills in patients with Parkinson's disease, *Brain*, 120, 1325-37.

Sonn U, Törnquist K and Svensson E. (1999) The ADL taxonomy – from individual categorical data to ordinal categorical data. *Scandinavian Journal of Occupational Therapy* 1999; 6, 11-20.

Stine OC, Pleasant N, Franz ML, Abbott MH, Folstein SE and Ross CA (1993) Correlation between the onset age of Huntington's disease and the length of the trinucleotide repeat in IT-15. *Human Molecular Genetics*, 2, 1547-1549.

Stuss DT and Benson DF (1984) *Neuropsychological studies of the frontal lobes*. New York, Raven Press.

Stuss DT and Benson DF (1986) *The frontal lobes*. New York, Raven Press.

Sulaiman S (2007) *Learning to live with Huntington's disease one family's story*, Jessica Kingsley Publishers, London and Philadelphia.

Soloman AC, Stout JC, Johnson SA, Langbehn DR, Aylward EH, Brandt J, Ross CA, Beglinger L, Hayden MR, Kieburtz K, Kayson E, Julien-Baros E, Duff K, Guttman M, Nance M, Oakes D, Shoulson I, Penziner E and Paulsen JS (2007) Verbal episodic memory declines prior to diagnosis in Huntington's disease, *Neuropsychologia*, 45, 1767-1776.

Tibben A, Vegter-van der Vlis M, Skraastad MI, Frets PG, Van der Kamp JJP, Niermeijer MF, Van Ommen G-JB, Roos RAC, Rooijmans HGM, Stronks D, Verhage F (1992) DNA-testing for Huntington's disease in The Netherlands: A retrospective study on psychosocial effects, *American Journal of Medical Genetics*, 44(1), 94-99.

Törnquist K, Sonn U. (1994) Towards an ADL taxonomy for occupational therapists. *Scandinavian Journal of Occupational Therapy* 1994; 1: 69-76.

Travers E, Jones K, Nicol J (2007) Palliative care in Huntington's disease, *International Journal of Palliative Nursing*, 13(3), 125-30.

Trembath MK, Horton ZA, Tippett L, Hogg V, Collins VR, Churchyard A, Velakoulis D, Roxburgh R, and Delatycki MB (2010) A retrospective study of the impact of lifestyle on age at onset of Huntington's disease. *Movement Disorder*, 25(10), 1444-50.

Van Duijn E, Kingma EM, Van der Mast RC (2007) Psychopathology in verified Huntington's disease gene carriers, *Journal of Neuropsychiatry and Clinical Neuroscience* 19(4), 441-448.

Van Duijn E, Reedeker N, Giltay EJ, Roos RAC and Van der Mast RC (2010) Correlates of apathy in Huntington's disease, *Journal of Neuropsychiatry and clinical Neurosciences*, 22(3), 287-294)

Van Zomeren AH and Spikman JM (2008) The assessment of attentional impairments in Halligan and Wade (2008) *Effectiveness of rehabilitation for cognitive deficits* ch 7 Testing speed and control, Oxford University Press.

Ward J, Sheppard JM, Shpritz B, Margolis RL, Rosenblatt A and Brandt J (2006) A four year prospective study of cognitive functioning in Huntington's disease. *Journal of the International Neuropsychological Study* 12, 445-454.

Watson R and Fourie M (2004) *Occupation and occupational therapy*. In: Watson R and Schwartz L (eds) *Transformation through occupation*. Whurr, London, 19-32.

Watt DC and Sellar AS (1993) A clinico-genetic study of psychiatric disorder I Huntington's chorea. *Psychological medicine*. Monograph supplement 23, 1-46 in Harper PS (ed) (1996) *Major problems in neurology: Huntington's disease*, second edition, Saunders, London.

What is a lasting Power of Attorney? Directgov website, [http://www.direct.gov.uk/en/Governmentcitizensandrights/Mentalcapacityandthelaw/Mentalcapacityandplanningahead/DG\\_185921](http://www.direct.gov.uk/en/Governmentcitizensandrights/Mentalcapacityandthelaw/Mentalcapacityandplanningahead/DG_185921) accessed on 11/5/11 at 16.04

Wilcock A (1998) *Occupation for Health*. *British Journal of Occupational Therapy* 61(8): 340-345.

Williams JK, Skirton H, Paulsen JS, Tripp-Reimer T, Jarmon L, McGonigal Kenny M, Birrer E, Hennig BL and Honeyford J (2009) The emotional experiences of family carers in Huntington's disease, *Journal of Advanced Nursing*, Jan 09, 789-798.

Skilbeck J and Pavne S (2005) End of life care: a discussion analysis of specialist palliative care nursing. *Journal of Adv Nurs* 51(4): 325-34 in Travers E, Jones K, Nicol J (2007) *Palliative care in Huntington's disease*, 13(3), 125-30.

Sweetland J, Riazi A, Cano SJ and Playford ED (2007) Vocational rehabilitation services for people with multiple sclerosis: what patients want from clinicians and employers. *Multiple Sclerosis*, 13(9), 1183-1189.

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

World Health Organization. Palliative care What is it? accessed at <http://www.who.int/hiv/topics/palliative/PalliativeCare/en/> on 14.3.11

**Production of the Guidelines**

This guideline has been developed by the European Huntington's Disease Network (EHDN) Standards of Care Occupational Therapy Group, a group of collaborating Occupational Therapist and health care professionals from across Europe and the USA. The group was chaired by Dr Sheila Simpson.

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