Huntington's Disease Association

Treating movement disorders in people living with Huntington's disease

EHDN European Huntington's Disease Network

Advancing Research, Conducting Trials, Improving Care

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Contents:

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Introduction

Movement disorders are one of the most widely known and visible symptoms of Huntington's disease, and although people with Huntington's disease will often experience cognitive or behavioural symptoms before motor symptoms, the presence of motor symptoms is still currently used as a formal indication of disease onset.

Due to the progressive nature of Huntington's, symptoms will change constantly over time. It is therefore important to re-evaluate treatments. Sometimes, the most helpful thing a clinician can do is discontinue an unnecessary drug. People living with Huntington's disease can be highly vulnerable to side effects, therefore starting on low doses and increasing slowly is recommended.

Polypharmacy should be avoided where possible, and non-pharmacological interventions can also be helpful.

Multi-disciplinary care

The complex nature of Huntington's disease means that it is essential to provide multi-disciplinary team care.

Neurologists, psychiatrists, psychologists, physiotherapists, occupational therapists, speech and language therapists, dieticians, nursing and social workers should all be involved in providing assessment and treatment.

Uncontrollable, abnormal and involuntary movements (chorea)

Most people living with Huntington's disease develop uncontrollable, abnormal and involuntary movements (chorea), which affect the trunk, face and limbs.

Discuss with the person with Huntington's if their movements are causing them problems or distress, as often they are not. Caregivers will often be more aware of, and more distressed by, the involuntary movements than the person with Huntington's disease. Be aware that hunger, infections, pain and medication side effects, can potentially worsen chorea.

Treating chorea with pharmacological agents can often affect other aspects of the movement disorder and can have a detrimental effect on cognition and mood. Consider non-pharmacological interventions such as massage. Although chorea may worsen in the early to middle stages of the disease, it generally lessens with more advanced disease, so reassessment of medication and reducing or withdrawing it may be necessary.

Although tetrabenazine is one of the few drugs for which there is clinical trial data on its effects on chorea, its administration can also be associated with cognitive changes and depression. It should therefore be avoided in anyone with current depression or a history of serious depression. Moreover, the starting dose in the National British Formulary is given as 25mg tds, but this is too high for many people with Huntington's disease. Consider starting with half this dose or less and to increase only if necessary.

In the UK, it is common amongst Huntington's disease specialists to use second-generation neuroleptics as a first-line treatment. This can be particularly useful when a person has a related mental illness or behavioural disorder.

Using a single drug to treat chorea is preferred because combination therapy increases the risk of adverse effects and may complicate the management of non-motor symptoms. As the disease progresses, chorea often subsides, and the Parkinsonian features of Huntington's disease become more prominent. Therefore, it is important to monitor this to ensure that someone is not receiving treatment unnecessarily.

A risk assessment should take place to identify whether protective measures need to be taken to avoid traumatic injury or choking, such as during mealtimes. This can occur due to progressive muscle weakness and poor coordination affecting the face, neck, and swallowing muscles. Occupational therapists and physiotherapists can advise on equipment, adaptions, and positioning techniques.

Young people living with Juvenile Huntington's disease are less likely to experience chorea, but are more likely to have muscle contractions and stiffness.

Involuntary muscle contractions and spasms (dystonia)

Dystonia is a neurological disorder that causes involuntary muscle contractions and spasms, resulting in abnormal postures, twisting, and repetitive movements. It can affect the entire body or just a specific part.

Dystonia refers to abnormal postures that can affect all parts of the body. People's experience of dystonia can range from a slight, intermittent, abnormal posture to severe muscle twitching, which significantly affects movements and day-to-day life.

Physiotherapy can help to maintain the range of movement and prevent the development of contractures.

Botulinum Toxin (Botox) injections can be helpful in some patients who have muscle stiffness and spasticity, particularly when this is most marked in a single or a small number of locations. A referral should be made to a specialist botox clinic for assessment.

Sitting is often a major problem for people with Huntington's disease, as people are at risk of slipping out of a conventional chair because of involuntary twisting and arching movements. Customised chairs can therefore be helpful. People should always have an assessment by an occupational therapist, even if they are buying a chair themselves.

Rigidity

Muscles are likely to become stiff and rigid. This is caused by an increase in muscle tone, leading to resistance to passive movement, which leads to

stiff joints and limited range of motion.

The use of neuroleptics or tetrabenazine may cause or increase rigidity. Consider reducing dosage or ending the use of these drugs if rigidity is causing a person distress.

This decision would need to be made by considering whether the overall benefits of these drugs in treating chorea and / or behavioural symptoms outweigh the distress caused by increased rigidity.

The place of Levodopa is unclear and controversial. However, there are anecdotal reports of Levodopa providing partial and temporary relief of the akinetic–rigid symptoms of Huntington's disease, especially in juvenile forms. If tried, treatment with levodopa should be started gradually, and the total daily dose is usually lower than in Parkinson's disease.

Physiotherapy may improve or maintain mobility and prevent the development of contractures and joint deformity.

Restlessness (akathisia)

Akathisia refers to an unpleasant feeling of restlessness that leads to a person being unable to sit still. If a person is experiencing akathisia, then a priority is to identify whether it is caused by a side effect of medication.

Tetrabenazine, neuroleptics and selective serotonin reuptake inhibitors (SSRI) may cause akathisia in Huntington's disease and reducing the dose or changing the treatment may be helpful.

Sudden muscle contractions (myoclonus)

Myoclonus refers to sudden, often painful, shock-like involuntary contractions of a muscle or group of muscles. It is more common in Juvenile Huntington's disease, where it may be mistaken for a seizure.

If myoclonus is impacting the functional capacity of a person with Huntington's disease, then treatment with sodium valproate or clonazepam,

used alone or in combination, and in escalating doses, is recommended.

Levetiracetam is a therapeutic alternative. If myoclonus is not associated with epileptic seizures, piracetam is an alternative.

Benzodiazepines, in particular clonazepam, can be used to manage myoclonus. This decision will need to be informed by considering the adverse effects of benzodiazepines. These include sleepiness, an increased risk of falls, and drug dependency.

Excess salivation

Many people with advanced Huntington's disease experience problems with excess salivation and drooling. This can be distressing for them and cause difficulties swallowing when saliva pools in the mouth. Drooling can result in sores to the mouth and chin and make eating and drinking more difficult.

Non-pharmacological interventions include postural management, supports for the head and neck and manually assisted cough techniques.

Medications such as transdermal hyoscine (scopoderm patches), sublingual atropine 0.5% drops, glycopyrrolate, and ipratropium bromide inhalers can be helpful. Palliative care specialists can further advise on these and on appropriate doses. However, people should be closely monitored to ensure that their mouths, mucous membranes and skin do not become too dry.

In cases where medication is not effective, botox injections may be helpful, and a referral to a specialist botox clinic could be considered. For people who have thick secretions, it is important, where possible, to ensure that they are well hydrated, and humidifiers, steam inhalation and nebulisers can help.

Difficulty walking or keeping posutre

Difficulties in walking and maintaining balance develop due to the progression of movement disorders in Huntington's disease. People struggle to regulate the number of steps they walk, experience variability in step width and length, and make more small, unconscious movements to maintain balance and stay upright (postural sway). Support should be

offered as early as possible and be continued and adapted throughout a person's life.

A wheelchair can be used for longer excursions, and other assistive devices, such as a four-wheeled walker, for shorter distances, or in the home. Walkers with wheels may be particularly useful when rigidity or loss of balance is a problem. People who are particularly prone to falls sometimes wear helmets, or elbow and knee pads, to minimise injury.

Working on core strength can improve balance, because good core stability is required to ensure safe and effective movement at the hip, knee, and ankle. Physiotherapists can provide guidance on exercises to improve core strength. Physiotherapy may also help by teaching people how to minimise injury in a fall and how to get up again after a fall. Further information can be found in our physiotherapy guideline.

Bruxism

Bruxism is an involuntary clenching with excessive contraction of the jaw muscles. It typically causes lateral movements (or front to back) responsible for gnashing, and can lead to tooth damage.

Injecting botox into the masseter muscles is a recommended initial treatment for bruxism. Customised protective mouth guards may be helpful, mostly in early-stage patients, but should be assessed on a case-by-case basis. Consider a referral to a dental specialist who may be able to make a shield which can provide tooth protection.

Bruxism may occur as a side effect of neuroleptics and selective serotonin reuptake inhibitors (SSRIs), therefore reducing their dose should be considered.

Manual Dexterity

A person's ability to use their hands in a skilful, coordinated way to grasp and manipulate objects can be impaired due to chorea / dystonia / akinesia / rigidity. However, it can also occur without these symptoms being present.

Neuroleptics and tetrabenazine may have a small beneficial effect on dexterity due to reducing chorea. However, they may also have a detrimental effect on dexterity by contributing to slow or difficult movement, or a lack of movement (bradykinesia).

Management with physiotherapy and occupational therapy may be useful to reduce the functional impact of fine motor skill deterioration. Adaptive aids may help to compensate for the deterioration of manual dexterity.

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Resources

Huntington's Disease Association resources

For professionals

The Huntington's Disease Association offers a range of resources to support professionals working with people affected by Huntington's disease.

You can find our resources at **www.hda.org.uk**

Join our professional-only mailing list to receive updates on events, webinars, and new resources tailored to your needs. You can sign-up at **www.hda.org.uk/professionals-and-training/**

Support for children and young people

Find information on our Huntington's Disease Youth Engagement Service (HDYES) by searching for "HDYES" on the website.

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