Nutrition and Huntington's Disease

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Huntington's disease, previously known as Huntington's Chorea, is a hereditary disorder of the central nervous system, resulting from a 'faulty gene' known as 'HTT' on chromosome four. Huntington's disease causes damage to the nerve cells in the brain, primarily in the basal ganglia, thalamus and cerebral cortex. There is a 50 per cent chance of inheriting Huntington's disease if one parent carries the faulty gene.[1, 2] Huntington's disease causes gradual physical, cognitive and emotional changes, with the onset of symptoms normally occurring between the ages of thirty to fifty years.[1] Weight loss is common in Huntington's disease, especially as the disease progresses,[2-5] with cachexia noted in the end stages of the disease.[6] At present there is no cure for Huntington's disease, the primary aim of medication is symptom management.[1]

Nutritional screening

Initial nutritional screening into services provided, including visits to day centres or routine clinic appointments, as well as on admission to acute or mental health hospitals, should include the use of a validated screening tool such as the 'Malnutrition Universal Screening Tool' ('MUST').[2, 7] In addition to this, frequent monitoring of the patient's weight and oral intake is recommended due to the risk of rapid weight loss even when patients are identified as being 'low risk'.[2] Where dietetic services are stretched training could be offered to support monitoring undertaken by members of the multi-disciplinary team, triggering a referral on to the dietitian for further support and individualised care planning where there is cause for concern.

Nutritional assessment

Patients with Huntington's disease are known to have a lower body mass index than matched controls;[2, 4, 8] this becomes more marked as the disease progresses.[8] The cause of weight loss in Huntington's disease is not fully understood and may be due to a combination of factors such as increased energy expenditure, metabolic changes and reduced oral intake.[2, 9] A potential cause of weight loss may be increased involuntary movements, causing increased energy expenditure.[6, 3, 5, 2] Weight loss may also be caused by difficulty experienced controlling voluntary motor skills to physically prepare meals, or to bring the food from the plate to their mouth, resulting in a reduced oral intake.[9, 10] Anxiety is known to further exacerbate the involuntary movements of Huntington's disease.[11] As the experience of eating becomes more difficult this may cause increased anxiety, which in turn can result in increased involuntary movements thus causing further difficulty and anxiety – a vicious cycle – ultimately causing a reduction in oral intake.[10] Nutritional intake has been found to be highly variable in the more advanced stages of Huntington's disease,[5] which may be due to an increase in involuntary movements and loss of voluntary motor control as the disease progresses.

Nutritional assessment undertaken by the dietitian should include weight history, a full diet history, taking into consideration intake of vitamins and minerals.[2] It is important to note that when patients with Huntington's disease are classified as overweight it may not be appropriate to provide weight loss advice due to the risk of rapid weight loss and deterioration as the disease progresses.[2] The European Huntington's Disease Network Dietitians Standards of Care Group[2] recommend that patients with Huntington's disease aim for a body mass index of between 23-25 kg/m^2. It is recommended that standard energy equations are used to calculate nutritional requirements. However, dietitians should be mindful that patients with Huntington's disease may have much higher calorie requirements than those estimated. Therefore, regular monitoring and dietetic review as part of the multi-disciplinary team is vital. The aim of dietetic input should be to maintain a healthy weight, optimise oral intake and prevent weight loss. The European Huntington's Disease Network Dietitians Standards of Care Group recommendations for nutritional requirements are shown in **Figure 1**.[2]

Figure 1: Summary of Nutritional Recommendations from the European Huntington's Disease Network Standards of Care Dietitians Group[2]

- 25-35Kcal/Kg/day
- 0.8-1.5g protein/kg/day
- Fat and carbohydrate as general population.

Oral nutritional support

The patient and carers/family members should be involved in the care planning process, promoting continued independence and dignity.[12] Care plans including nutrition support measures are appropriate for use with patients with Huntington's disease, this may include the use of food fortification and increasing nourishing snacks and drinks. In addition, where these measures are not sufficient to maintain weight and nutritional status, the use of oral nutritional supplements should be considered, taking into account the patients swallowing ability.[13, 2] A small study of the use of oral nutritional supplements in patients with Huntington's disease found that provision of two supplement drinks per day providing a total of 473kcal, over ninety days, was effective in promoting weight gain, increasing mid upper arm circumference and percentage body fat in over 68 per cent of participants in the study.[13]

Oral care

There is an increased presentation of patients with Huntington's disease to dental services with tooth decay; this may be due to difficulties holding a tooth brush or loss of drive to maintain oral care. Patients with Huntington's disease may find visits to the dentist particularly difficult due to the need to sit still for treatment. It is, therefore, essential for dietitians and the multi-disciplinary team to promote the need for good oral care to both patient and family/carers.[2, 10]

Dysphagia

Dysphagia is common in Huntington's disease and may become worse as the disease progresses.[2, 10, 14] Dysphagia increases the risk of aspiration pneumonia.[14] A recent European retrospective study looking at causes of death in Huntingdon's disease, found that pneumonia was the most frequent cause of death in their participants, causing over 25 per cent of deaths in the study.[15] Therefore, it is important that patients with Huntington's disease have their swallow regularly assessed by a speech and language therapist to reduce this risk. Signs and symptoms of dysphagia are shown in **Figure 2**. It is also important for members of the multi-disciplinary team to support the recommendations made by speech and language therapists providing practical advice and support to patients and carers to adapt to a texture modified diet.

Figure 2: Signs and Symptoms of Dysphagia[10]

- Gurgly/husky voice after eating or drinking
- Repeated swallow
- Coughing/spluttering
- Food/drink spilling from mouth
- Food pooling in the side of the cheek
- Tiring easily when eating
- Increased anxiety/frustration at mealtimes
- Weight loss
- Chest infections.

Patients with Huntington's disease may appear to be always hungry, 'wolfing' food down at a considerable rate, even when larger portions or more frequent meals are provided. This can increase the risk of aspiration. At mealtimes carers should prompt the patient to slow down and take their time.[10]

Enteral feeding

Where possible non oral feeding, such as use of a percutaneous endoscopic gastrostomy (PEG), should be discussed with the patient and their family or carer in the early stages of Huntington's disease in order to establish their wishes in the case of swallow deterioration as the disease progresses, when communication may be more difficult and capacity to make this decision diminished.[2] Any decision or expression of the patient's wishes about non oral feeding should be clearly documented by the multi-disciplinary team in the patients records to refer to for future use. If the patient has made an advance directive regarding non oral feeding this should also be documented and respected by law.[2]

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Cognitive and behavioural changes

Huntington's disease causes changes to the subcortex and its connections to the front of the brain, which lead to changes in behaviour, in particular causing mental rigidity, emotional blunting, loss of drive and initiative. It should also be noted that some changes in behaviour may be caused by frustration, with loss of independence experienced as the disease progresses.[11, 16] As patients with Huntington's disease exhibit changes in behaviour this can cause significant upset and anxiety for family members and carers, who may perceive the person to be becoming lazy or awkward.[11, 16] Maintaining a structured routine may help the patient with Huntington's disease to maintain some independence and maintain dignity, for example, by including personal hygiene tasks as part of the daily structure. Meal and snack times should also be built into the daily routine, having set meal times may optimise oral intake and reduce carer anxiety around when to offer food and drink.[16]

Patients with Huntington's disease often lose the ability to multi-task in the early stages of the disease, such as being able to watch television and eat or hold a conversation and walk.[16] Therefore it is important that dietary strategies to promote oral intake include assessment and consideration of the mealtime environment. This may include working with families or carers to minimise distractions and prevent the patient from becoming over whelmed by distractions,[9] such as turning the television off at mealtimes or not having conversations whilst the person with Huntington's disease is eating or drinking. An assessment by an occupational therapist should also be considered to support with the activity of eating and drinking, providing specialist aids or seating, to promote continued independence for as long as possible. The mealtime environment should also be clean, pleasant and as calm as possible. However, keeping the mealtime experience calm may be a considerable challenge for the family/carers of patients with Huntington's disease who may be worried about them not eating or drinking sufficient amounts.[10, 11] It is important that the multi-disciplinary team continue to provide support for family and carers, and that dietary strategies are individualised to optimise oral intake. This may also involve changing mealtimes to the time of day when the patient is at their most awake and alert, whilst maintaining a structured routine.

Communication

Communication may become an increasing challenge in Huntington's disease, with many patients being unable to verbally communicate by the end stages of the disease.[17] It is vital, as professionals, to work with families and carers who are experts in caring for their relative to aid interpretation of non-verbal cues, such as facial expression, when undertaking dietetic assessment. Multi-disciplinary working with speech and language therapists to develop and utilise effective communication tools is essential.[17]

Mental health

Depression is common in Huntington's disease, especially in the early stages of the disease, which may be due to the adjustment to a progressive disease as well as changes in the brain.[11, 18] Depression can adversely affect oral intake, so it is important that this is taken into consideration when completing a nutritional assessment and developing dietary strategies to optimise oral intake. Obsessive compulsive behaviour is also common in Huntington's disease[19] and may impact on food choices.[2] Dietary strategies and care planning should take this into consideration whilst working closely with the multi-disciplinary team.

Summary

Patients with Huntington's disease are at risk of weight loss, especially as the disease progresses, although the causes for this are not yet fully understood. Nutritional intake in Huntington's disease can be affected by involuntary movements, dysphagia, and cognitive and behavioural changes. The dietitian is a vital part of the multi-disciplinary team throughout all stages of Huntington's disease, providing advice and support to optimise oral intake and maintain a healthy weight.

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