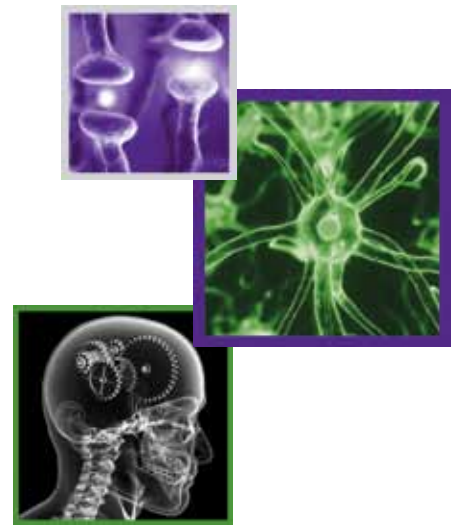
SPECIAL REPORT

Nutritional management of individuals with Huntington's disease: nutritional guidelines



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Practice Points

- Good nutritional care is a fundamental element of the management of individuals with Huntington's disease.
- Nutritional assessment and care planning for individuals with Huntington's disease must take account of the stage of the disease and feeding difficulties individuals may experience.
- Patient's weight must be monitored regularly and kilocalories should be altered in a way that is acceptable to the individual, until an agreed target weight is achieved.
- Due to the variability in energy requirements and the potential for rapid weight loss, early individual assessment and regular reviewing of nutritional care plans are vital.
- Many individuals have very increased energy requirements and it is essentials to provide adequate macro- and micro-nutrients.

SUMMARY The delivery of good nutritional care is a fundamental element of the management of individuals with Huntington 's disease and all patients with Huntington's disease will, at some time, need dietary intervention because of the sequela of the disease, yet there are no European nutritional guidelines. The European Huntington's disease network Standards of Care Dietitians group has brought together expert Dietitians from across Europe to produce nutritional guidelines to improve the nutritional management of individuals with Huntington's disease. The guidelines were developed to promote optimal nutritional screening, assessment and management of individuals throughout all stages of the disease, with the aim of improving the standard of nutritional care delivered. Literature was systematically searched in an attempt to ensure that the recommendations are based on sound evidence and where evidence is lacking; specific guidance is based on consensus expert dietetic opinion. The provision of nutritional care varies widely between countries. Implementation of these nutritional guidelines across Europe should improve the quality of nutritional care delivered to individuals with Huntington's disease.

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Future Medicine part of fsg

Huntington's disease (HD) is a complex, inherited neurodegenerative disorder characterized by a progressive movement disorder, cognitive impairments, personality and psychiatric problems, as well as metabolic changes (Box 1) [1]. It is caused by an unstable expansion of a CAG sequence within the Huntingtin, known as *HTT*, gene which is located on chromosome 4 [2]. The protein encoded by the *HTT* gene is important in normal brain and neuronal development. The expanded CAG sequence leads to the production of an abnormal protein that causes brain cell dysfunction and ultimately neuronal cell death primarily in the basal ganglia but also the thalamus and cerebral cortex [3]. This single gene disease follows an autosomal, dominant inheritance affecting both men and women. Each offspring of an affected parent has a 50% chance of inheriting the gene mutation and developing the disease. Symptom onset usually occurs in mid age but a juvenile onset (<20 years of age) and late-age onset are also recognized. To some extent the age of onset is related to the size of the mutation [4].

Box 1. Symptoms of Huntington's disease.

Motor symptoms
- Chorea
- Dystonia
- Rigidity
- Bradykinesia
- Gait disorder

Cognitive changes
- Difficulties planning and organizing
- Lack of initiation (activities and conversations)
- Perseveration
- Impulsivity
- Irritability and temper outburst
- Perceptual problems
- Unawareness/ Lack of Insight
- Difficulty in new learning

Speech, language and communication problems
- Specified in text

Neuropsychiatric/behavioral symptoms
- Depression
- Obsessive–compulsive disorder
- Irritability
- Apathy
- Anxiety

Other
- Metabolic changes
- Sleep disturbances
- Dysphagia

The prevalence of HD in Europe is estimated to be approximately four to ten individuals per 100,000. However, recent reports suggest that the number of individuals with HD is underestimated and that the true prevalence is considerably higher [5].

HD is a family disease and most individuals have seen one of their parents of other family member go through the stages of the disease. This makes them very aware of what will or might happen to them in the future and has a significant influence on their knowledge of the disease and decision making, for example consenting to enteral tube feeding.

Stages of HD

There have been several attempts to define the stages of progression of HD and once symptomatic stages are currently based upon the individuals total functional capacity (TFC) score [6]. Current research may provide further clarification [7]. For these guidelines nutritional recommendations have been made in three stages of the disease; early, mid and late.

Early-stage disease cannot be defined as the time of diagnosis since diagnoses can be made at variable times. Most individuals present with early neurological features such as subtle changes in coordination and mild involuntary movements [3]. Psychiatric symptoms and cognitive changes often occur prior to the development of these motor signs. Hence in early stages depression, irritability, difficulty in planning and organizing and prioritizing activities can be experienced. Although maybe slightly less functional, people remain largely independent in their daily activities and lives [8].

Mid-stage disease would be deemed to have been reached when the affected person has to cease work, or at least change employment to a post less challenging [8]. Involuntary movements increase and voluntary tasks will become more difficult. Executive dysfunction, including organizing, reasoned decision making, memory difficulties and reduced ability to learn new information make cognitive tasks more difficult [3].

Late-stage disease occurs when employment becomes impossible, the individual is no longer able to live independently [8], self-care ceases and cognitive decline is obvious. At this stage patients are severely limited in their mobility and are often chair or bed bound. Some patients maintain the ability to feed themselves to a late

stage, but swallowing difficulties and involuntary movements create increasing difficulties. Cognitive deficits progress to a global dementia.

Weight loss is a common and recognized feature throughout all stages of the disease [9–11] and its etiology remains uncertain. Its progressive nature is often marked in the later stages [9,12,13] of the disease and correlates with the CAG repeat number [14]. Evidence, including animal studies, suggests that there are several possible contributing factors, such as:

- Increased metabolic rate and higher energy expenditure perhaps secondary to hypothalamic dysfunction [10,15]
- Possible peripheral abnormalities caused by the expression of the faulty protein in those tissues [12]
- Insufficient intake due to swallowing and feeding difficulties [16–18], including xerostomia [19]
- Hyperkinetic movement disorder [16]
- Possibly a combination of all above points

There is great variation in the presence of symptoms and the impact of these on an individual's nutritional intake and status. Although within this standards of care document, nutritional care has been subdivided into these three stages, according to the usual prevalence of the onset of the difficulty, it is important to note that individuals may experience the difficulties at any time of the disease progression.

Role of the European Huntington's Disease Network

The European Huntington's Disease Network (EHDN) provides a platform for clinicians and families to work together to find a cure for HD, to co-ordinate research and in the meantime to elicit best practice for the care of individuals with HD. Working groups were created to address the various themes (http://www.euro-hd.net/html/network/groups, 2010); one group focusing on Standards of Care (SoC) with specialist subgroups. Care provision of HD varies widely between countries. The SoC working party has produced a consensus view for management of HD, which reflects the opinions of experts in their field, and will enable others to provide best care for the HD patient and their family.

The EHDN Standards of Care Dietitians group has brought together expert dietitians from across Europe to produce nutritional standards of care to improve the nutritional management of individuals with HD. The information contained within this guidance is based on the available evidence in the nutritional care of individuals with HD, current nutritional support guidance and consensus of dietetic opinion within the field.

Aims of the nutritional guidelines

The aims of the Nutritional Management of Individuals with HD guidelines are to provide nutritional guidelines for dietitians to allow for optimal nutritional screening, assessment and management of individuals throughout all stages of HD, with the aim of improving the standard of nutritional care delivered and to ensure the nutritional management of individuals with HD is based on sound evidence and current best clinical practice.

Nutrition guideline development

The delivery of good nutritional care is a fundamental element of the management of individuals with HD. This guidance attempts to bring together the research evidence and expert dietetic opinion from across Europe in making nutritional recommendations for individuals with HD. However, it is important to acknowledge the differences in approaches between Countries and professionals must also take account of their national legal requirements, national and local policies and procedures and professional codes of conduct in the interpretation of this guidance. Prior to the development of this guideline the literature was systematically searched in an attempt to ensure that the recommendations are based on sound evidence. However, a number of challenges were faced as the published literature in the field is limited, especially in terms of methods of nutritional screening and quantifying nutritional requirements in this patient group. The HD nutritional evidence base was difficult to interpret for clinical practice for the following reasons:

- Few studies provided robust evidence for nutritional care in HD
- Studies had small sample sizes
- An extensive range of outcome measures were used and these were not directly comparable
- Large gaps in the evidence base

- Inconsistencies in definitions, for example, malnutrition

Although the evidence was graded in accordance with the SIGN guidelines [20], all the specific nutritional recommendations contained in this Guideline are based on consensus of the expert dietetic opinion of the EHDN Standards of Care Dietitians Group, informed by the generic nutritional guidelines and established clinical practice, because of a lack of evidence in the field.

Literature review

A systematic literature search was performed electronically using OVID Medline (1988–2009), OVID Embase (1988–2009) and EBSCO Cinahl (1988–2009) to evaluate evidence on BMI, weight loss, energy requirements and recommendations for nutritional support. A search strategy was developed in collaboration with the members of the Standards of Care Dietitians Group. The literature search was updated during the development of the guideline. All relevant publications were identified and categorized to evidence statements according to SIGN methodology [20]. Due to the lack of scientific evidence recommendations were primarily formulated based upon clinical experience and expert consensus from within the EHDN SoC Dietitians Group.

The importance of nutrition in HD

Key nutritional priorities

Recommendations within published, generic, evidence-based nutritional guidelines [21,22] are generally applicable in HD. However, due to the cognitive, motor, psychiatric and communication problems which occur in this progressive, degenerative condition additional nutritional factors need to be considered.

Unintended and sometimes rapid weight loss is common in people with HD and often occurs despite a good appetite [10,23] and a high calorie intake [10]. Observation has shown that there can be variations in energy requirements throughout the course of the disease and between HD individuals [10,11] but generally speaking, in the premanifest and early stages of HD patients may already have higher energy requirements than the normal population [10,23,24]. This may explain why their BMI is often lower compared with the normal population. A higher bodyweight at diagnosis has been associated with a slower progression of the disease [25]. The cause of increased energy requirements and weight loss is not clear and most likely to be multifactorial. However, due to the variability in energy requirements and the potential for rapid weight loss, early individual assessment and regular reviewing of nutritional care plans are vital. Therefore, the EHDN SoC Dietitians Group recommend that, on diagnosis, individuals with HD should receive the following dietetic care.

Ensuring adequate nutritional intake throughout each stage of the disease

This will require adequate provision of suitable foods and fluids, either by the individual or their carers. Distinct nutritional challenges at each stage of the disease require careful dietetic management. Many individuals have very increased energy requirements and it is essentials to provide adequate macro- and micro-nutrients.

When weight loss occurs it can be very rapid therefore preventing weight loss and maintaining a healthy weight is very important. However this can be difficult to achieve because the symptoms of HD also often have a profound effect on food intake.

Protective weight management

There is a high risk of rapid weight loss for many individuals but for some, rapid weight gain may occur. Studies have found that individuals with HD have a lower BMI than controls at diagnosis [14,24] and younger age of onset usually denotes more aggressive disease and possibly an even lower BMI [25]. Rapid weight loss can result in loss of muscle mass, weakness, apathy, depression, susceptibility to infection and compromised chest status.

The risks may change during the progression of the condition but both unmanaged weight loss or weight gain will be problematic. For this reason weight should be carefully managed as standard weight management advice may be inappropriate for individuals with HD.

Nutrition & function

Poor nutrition is known to impact on function in certain neurological populations, for example, elderly, stroke patients [26,27]. Although there are no studies looking at nutrition and function in HD *per se*, due to the physical and cognition difficulties poor nutrition has a much greater impact on this group. The affected functions include strength and muscle mass, levels of

fatigue; already low in this population group, mood (which can already be compromised by the disease itself) and cognitive function. It therefore seems reasonable to ensure optimal nutrition so that these factors are not further exacerbated by poor diet

Physical impact on nutritional intake

The main physical problems are difficulties with motor coordination and muscle control, which may result in uncontrolled movements and difficulty walking, speaking and swallowing. Uncoordinated movements are a challenge when handling cutlery and taking food to the mouth. Chewing and swallowing difficulties arise from uncoordinated movement of the face, neck, lips and diaphragm. This can make eating and drinking a tiring experience. Other common problems are vomiting, reflux and regurgitation of food as the systems for moving food through the digestive system become disrupted.

Cognitive changes affecting food intake

Cognitive changes which are to do with the ability to think, impact on problem solving, planning and sequencing of ideas. This includes the decisions before the food enters the mouth and may lead to an inappropriate food choice and/or eating too much too quickly. Concentration and memory can also be affected. Overall, an individual's awareness and ability to make judgments becomes more impaired.

Nutrition & emotion

Mood swings and changes in emotional responses are common, all of which can adversely affect behaviour. Depression can occur and affect the person's appetite and anxiety is common. Anxiety can make movements more pronounced and affect the swallowing process. Frustration may occur when faced with being unable to prepare food and eat in the usual way. Many people with HD have a great resistance to adaptive changes to their lifestyle which can make accepting advice for safer eating or altering the texture of food and fluid difficult.

Nutrition & oral health

Particular attention should be given to oral health especially where dietary advice increases the intake of simple carbohydrates and/or frequency of meals and snacks as dental decay can result. Due to their facial and body movements, they are a very difficult group for dentists to treat. Oral health will impact on the risk of chest infections from aspiration of bacteria-laden saliva and will also impact the ability eat and drink if the patient has dental problems or pain.

Multidisciplinary approach

Healthcare professionals should ensure that all people who need nutrition support receive coordinated care from a multidisciplinary team (MDT) [22]. It is recognized that members of the MDT will vary depending upon the setting and local policy, but we stress, the importance of management of specific difficulties by trained therapists, for instance, management of swallowing difficulties by appropriately trained speech and language therapists. Ideally, the team will also include those who have specialist skills in positioning and aides for feeding. HD is neurological condition of long duration and as such may have a lengthy palliative phase. Diagnosis of HD will have a major impact on the quality of life of the affected individual and their family [28]. A multidisciplinary approach is vital to ameliorate the problems encountered in caring for this population [29].

Behavioral aspects of the disease present major nutritional challenges

The disease presents a range of behavioral issues as a consequence of psychological and cognitive deterioration that can impact on nutritional intake. There may be an inability to express food preferences or to inform carers of discomfort when eating, leading to frustration which can present as challenging behavior. This can be especially apparent at mealtimes and requires skilled feeding techniques and careful management by carers. Obsessive behaviors, often seen in HD, can impact on food choices and dealing with these issues can be very challenging.

Respecting autonomy

Individuals with HD often lack insight into their own disease progression. They may not be aware of, or are not able to, acknowledge progressive symptoms. Individuals will often refuse advice and interventions because of a decline in cognitive ability, and especially in executive function. All nutritional advice provided should be set within the context of quality of life (QoL) and patient choice.

Table 1. Nutritional challenges and recommended standards of nutritional care for individuals with early stage disease.

	Nutritional challenge/recommended level of nutritional care	Rationale for the recommendation
1	Early assessment of nutritional intake and status	Severe rapid weight loss is problematic, although not universal An early assessment of nutritional intake is essential to the delivery of effective nutritional care Early dietetic assessment facilitates delivery of good nutritional care in later stages of the disease when the patient becomes unable to communicate effectively.
2	The baseline assessment should include weight, height, BMI, completion of a validated nutritional screening tool, calculation of percentage weight loss, record of usual weight. If unable to obtain weight, use alternative measurements such as mid-arm circumference	Nutritional assessment should be carried out by a trained health professional The nutritional status of individuals with HD can deteriorate rapidly
3	Ensure adequate nutrient intake including sufficient energy, protein, fluid, electrolytes, minerals, micronutrients and fiber needs (NICE, 2006), taking account of any increased nutritional needs	Some individuals have higher energy requirements [11]. Individuals should aim for a balanced intake of fat, protein and carbohydrates to provide the additional calories to maintain weight Excessive and frequent intake of simple sugars should be avoided where possible as this may impact on dental health If the dietary assessment indicates an inadequate intake of vitamin and minerals, a vitamin and mineral supplement should be considered
4	Consider impact of medication on nutritional intake and status	Medications used in the treatment of HD to suppress chorea may affect an individual's weight and food intake. For example, some neuroleptics may cause raised triglycerides, glucose, dry mouth, constipation, weight gain, increased appetite (See Medication table in **supplementary material**).
5	Discuss tube feeding within MDT and identify a member of the team to discuss it with the patient focusing on potential benefits and burdens. Record the individuals wishes	Discussing tube feeding in the early stages of the disease before cognitive function diminishes and communication becomes difficult. Many individuals have witnessed family members experience similar issues and are aware of the impact of the advancing disease Discussions must be patient led; some patients will want to discuss feeding fully but others will want to avoid or defer decision making. Group discussions or education facilitated by the dietitian may be beneficial

HD: Huntington's disease; MDT: Multidisciplinary team.

Nutritional assessment & evaluation

■ Nutritional screening tools

All patients with HD will at some time need dietary intervention because of the sequela of the disease. On diagnosis they should be screened for malnutrition and risk of malnutrition by a trained healthcare professional using a validated screening tool. Suitable tools include the Malnutrition Universal Screening Tool (MUST) [30], Mini nutritional assessment [31] and the short nutritional assessment questionnaire [32]. Screening should be repeated according to clinical need and local policy as specified in the screening tool adopted. It must be stressed that individuals with HD who are not underweight, and therefore not identified as 'at risk' on screening, may have an inadequate intake of energy and nutrients because of factors such as poor food choices or excessive alcohol intakes, therefore it is important that a diet history is taken regardless of the MUST score/screening outcome.

■ Nutritional assessment & evaluation

The EHDN Dietitians group suggest the following recommendations for the nutritional management of individuals with HD, based on a consensus of dietetic expert opinion. NICE [22] and ESPEN [21] guidance should be used as a guide where appropriate but additional issues need to be considered.

Nutritional support should be considered in those who are malnourished, as defined by any of the following:

- Unintentional weight loss greater than 10% within the last 3–6 months
- A BMI of less than 20 kg/m^2 and unintentional weight loss greater than 5% within the last 3–6 months [22]
- A BMI of less than 20 kg/m^2 (expert opinion is to aim to maintain BMI 23–25)
- Nutritional support should also be considered

Table 2. Nutritional challenges and suggested standards of nutritional care for individuals with mid-stage disease.

	Nutritional challenges/recommended level of nutritional care	Rationale for the recommendation
1	Monitor weight/weight changes should be recorded at every clinic visit If target weight not achieved/ weight loss >5% re-refer to dietitian	Nutritional goal in mid stage is often to maintain a healthy weight (BMI: 23–25 is recommended by the group), ensuring adequacy of nutritional intake to prevent rapid weight loss NB: caution is required for individuals who are overweight as a weight reducing diet may be inappropriate if the person is at risk of future rapid weight loss and feeding difficulties
2	Monitoring nutritional intake Discussion of food preferences Use of supplements or food fortification	Difficulties with social aspects of eating may impact on nutritional intake Poor food choices may be important to address in order to motivate behavior change while patients are cognitively able to understand the reasons Particular attention should be paid to any difficulties experienced as a result of advancing disease such as problems caused by reduced mobility, dexterity, cognition, swallowing and behaviours associated with advancing HD such as food obsessions food cramming, spillage, fear of choking or apathy
3	Food texture modification may be indicated Texture modified diets will require fortification to increase calorific value and nutritional supplement may be required	Referral to the SLT team for assessment is recommended where clinically indicated Appropriate texture of food and drink is required to minimize the risk of aspiration
4	Altered eating behavioural issues, for example, excessive coffee drinking, food cramming, and fixations	Drug use, excessive alcohol intake or constant smoking may impact on nutritional intake Dietary advice and intervention should focus on practical ways to modify intake recommending alternatives where appropriate, for example, decaffeinated coffee
5	Consider specific problems impacting on food intake, for example, sleep difficulties, inability to eat normal portions sizes, constipation, unawareness of importance of nutrition and eating difficulties	Provision of individualized nutritional care plan and implementation of person centred appropriate solution focused strategies to overcome nutritional difficulties

HD: Huntington's disease.

for those individuals who struggle to maintain their bodyweight and those at risk of malnutrition, defined as individuals who have:

- A: Eaten little or nothing for more than 5 days and/or are likely to eat little or nothing for 5 days or longer
- B: Food intake below 50–75% in preceding week
- C: A poor absorptive capacity and/or high nutrient losses and/or increased nutritional needs from causes such as catabolism.

Expert dietetic opinion is also that weight loss should be considered in individuals with either BMI of 27–30 with co-morbidities or for those with a BMI of over 30 without co-morbidities. Individuals should only be advised to lose weight under the supervision of a dietitian. They will require review every 2–4 weeks because of the risks of rapid weight loss in some patients.

Optimizing nutritional intake in the different stages of HD

■ Nutritional management of individuals with HD

The nutritional assessment and nutritional care planning process should ensure that the total nutrient intake of individuals with HD provides sufficient energy, protein, fluid, electrolyte, mineral, micronutrients and fiber needs [22]. The assessment must take account of the possible increased nutritional needs, especially kilocalorie requirements which can, in some individuals, be very high.

Gaba [33] recommended HD specific equations for the calculation of energy though there is a lack of HD specific calculations for protein and fluid requirements. Nutritional requirements can be calculated using a standard equation (such as Schofield, Harris Benedict) and the individual's current kilocalorie intake and weight history should form an integral part of the baseline nutritional assessment to determine estimated requirements.

Frequent monitoring of nutritional intake, weight and weight changes is required and adjustments to nutritional intake will need to be made to achieve the identified nutritional goals.

As a baseline for calculating requirements, the following NICE guidance [22] is appropriate but Dietitians must be aware that some individuals will have much higher kilocalorie requirements and other nutritional needs. The EHDN Dietitians group make the following HD specific recommendations, incorporating NICE Guidance:

- 25–35 kcal/kg/day total energy (including that derived from protein)
- Ratio of fat and carbohydrate same as normal population
- 0.8–1.5 g protein (0.13–0.24 g nitrogen)/kg/day
- 30–35 ml fluid/kg (with allowance for any extra requirements, input of fluid from other sources and any additional losses) offers a baseline for calculating requirements but in some individuals fluid requirements may be higher so it is important to monitor intake and adjust fluid as required, paying particular attention to the losses associated with difficulties drinking
- Adequate electrolytes, minerals, micronutrients (allowing for any pre-existing deficits, excessive losses or increased demands) and fiber if appropriate. In HD there is no evidence that vitamin and mineral requirements are altered and caution is appropriate to avoid excess intake especially where a range of nutritional supplements are recommended to increase kilocalorie intake. Dietary advice and/or supplements may be required in individuals where dietary intake is low, for example due to restricted or modified diet where there can be significant losses in food processing or where there is need especially to ensure adequate Vitamin C, iron, calcium and Vitamin D (in the case of vitamin D, important for individuals who are confined to indoors)
- Patient's weight must be monitored regularly and kilocalories should be altered in a way that is acceptable to the individual, until an agreed target weight is achieved.
- Dietary advice regarding fibre should be individualised advice and based on symptoms.

Nutritional assessment and care planning for individuals for HD must take account of the stage of the disease and the feeding difficulties individuals may experience. As with all clinical conditions, there are features of the disease that may affect the individual's ability to eat and drink, and each stage of the disease presents key challenges. The Nutritional challenges and recommended level of nutritional care are summarised and presented for early-stage disease (Table 1), mid-stage disease (Table 2) and end-stage disease (Table 3). Table 4 outlines the issues to be considered in the decision-making process for enteral tube feeding.

Table 3. Nutritional challenges and suggested standards of nutritional care for individuals with late-stage disease.

	Nutritional challenges/recommended level of nutritional care	Rationale for the recommendation
1	In addition to the assessment outlined in mid stage: Texture of diet and usual oral intake Use of supplements or food fortification Specific difficulties with eating and drinking Level of independence with eating and drinking Usual bowel pattern	Ensure good communication between members of the MDT to meet patient's nutritional needs and to have a comprehensive overview of factors impacting on ability to eat and drink Constipation can cause food refusal or/and increase risk of reflux or vomiting Difficulties with social aspects of eating may impact on nutritional intake. Particular attention to any difficulties experienced as a result of advancing disease such as difficulties with reduced mobility, dexterity, cognition, swallowing and behaviours associated with advancing HD such as food cramming, spillage or apathy
2	Care settings: On admission: Commence food and drink record charts Establish frequency of weighing Observe the patient eating at a meal time Establish a suitable target weight Determine if advance directives (AD) exist Establish a person's capacity	Identify level of dietetic intervention required Observation at meals highlights all aspects impacting on a persons ability to eat and drink a nutritionally adequate diet A target weight ensures the dietitian and nursing team have clear goals regarding weight management

HD: Huntington's disease; MDT: Multidisciplinary team.

Conclusion

Nutritional screening and the delivery of good nutritional care are fundamental elements of the care of an individual with HD. Other factors which impact on food intake should also be carefully considered but are outside the remit of these nutritional standards. These include effective communication strategies, appropriate feeding techniques and the importance of gathering relevant information regarding food preferences in the early stages of the disease before cognition becomes so impaired that communication becomes difficult. Feeding and communication guidance for dietitians and carers have been developed to support these nutritional standards of care. These address practical management issues, recognizing that the best form of support is not always straightforward as difficulties often arise as a result of the interaction of several factors. The guideline has been created for the information and support of dietitians caring for individuals with HD in hospitals, care homes and the community. Some dietitians might only care for a small number of clients with HD and are unable to develop expertise in this area. This framework can be a reference point for their practice. For clinicians and physicians involved in HD management this guide will offer an invaluable insight into the role of the dietitian and present assessment and referral criteria

It should be noted that although the guidance has been written specifically for patients with HD, much of the information may also be valuable across other neuro-degenerative diseases. The EHDN SoC group plan to audit compliance to these standards of care across the network and are planning a quality improvement program to support units to deliver against these standards.

Future perspective

Improving the nutritional care of individual with HD is likely to become much more of a priority focus for healthcare systems globally. Parenteral and enteral nutrition societies and associations are lobbying Governments and health departments to ensure that nutritional care is given the priority focus that it deserves. There is an urgent need for further research into the nutritional care of patients with HD as the role of nutrition in the treatment of the disease remains poorly understood.

Table 4. Decision making for enteral tube feeding.

	Recommended level of nutritional care	Rationale for the recommendation
1	If the person has an Advanced Directive stating they do not want a feeding tube, this is always upheld.	This is legally binding in many European countries
2	Discuss the benefits and burdens of enteral feeding as early as possible and record the patients wishes, family members should be involved in these discussions where appropriate If the person has capacity to decide whether or not they would want a feeding tube, then they should be educated to make an informed choice. Everything that is necessary to facilitate an individual's understanding and retention of the information should be tried. If capacity is uncertain than an assessment of capacity should be carried out Ideally this should not be done at a time of crisis. These discussions should take place prior to the need for enteral feeding arising	Family members should to be included in discussions (where appropriate) as they will often be supporting the patient Consent is required for patients able to make decisions about their care for any medical procedure/ treatment Patients do change their minds so it is good practice to routinely ask at intervals if their decision still stands These expressed wishes and views count as verbal advance directives and should always be documented for future reference (i.e., when the patient may no longer be able to communicate)
3	If the patient lacks capacity a multidisciplinary discussion is essential and local and national policy must be followed	It is not possible to make a Europe-wide recommendation given the differences in legal and clinical systems
4	The following should be considered to ensure a best interest decision is made: Any previous expressed views on PEGs Verbal directives to family members/ carers Family views on whether they would want this Perceived quality of life; this is very subjective Swallowing difficulty, fear of choking, infection history, weight change and level of distress eating causes Benefit or burden of feeding	In making a best interest decision it is important to look at the current situation for the person as it is. It is also relevant to know how they were as a person and what they would likely do in this situation if they were able to make a decision. This is where it is important to consult staff and family that may have had input with the person when they expressed such views

Production of the guidelines

This guideline has been developed by the EHDN SoC Dietitians Group, a group of collaborating dietitians, nutritional specialists and healthcare professionals from across Europe. The group is chaired by Sheila Simpson and Daniela Rae.

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■ **Understanding patients individual energy requirements is crucial to the provision of a nutritionally adequate intake of kilocalories and the variation in requirements can be quite significant and must be accounted for in clinical practice.**

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