

## 'My Practice'

# Management of Huntington's disease: the role of high energy enteral feeds

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### Background

Frank\* was diagnosed with Huntington's disease at 26 years of age. Following his diagnosis, he lived in sheltered accommodation and suffered a series of falls and choking episodes due to a reduction in cognition, coordination, balance and muscle strength. In his mid-thirties, his condition deteriorated and he was suffering severe chorea movements; he developed a need for a higher level of daily support. Consequently, he moved to a specialist care unit where he was referred for a speech and language therapy (SLT) and dietetic intervention.

### Initial Assessment

At the initial dietetic assessment, Frank's weight was 64kg, with a BMI of 19.3kg/m<sup>2</sup> and a Malnutrition Universal Screening Tool ('MUST') score of 2, indicating a high risk of malnutrition. His swallowing problems were a long-standing issue and Frank has lost 8.5% of his body weight over a 6-month period.

Due to a lack of evidence and guidelines with regards to the stress factor associated with chorea movements in Huntington's disease, it was difficult to determine Frank's daily nutritional requirements. They were estimated by the dietitian at 2800kcal, including a predicted 60% stress factor as a starting point to account for the energy used for his chorea movements, as well as a 400kcal per day addition to allow for weight gain.

The speech and language therapy assessment revealed that Frank required a soft, moist diet. When drinking, he required supervision with thin fluids in single sips using a straw. His swallow function was noticeably worse on days when Frank was tired or ill. He would manage his risk of choking and aspiration by limiting

his oral and fluid intake in response to how he felt, therefore having a significant impact on his nutritional intake. Frank also experienced a reduced ability to seal his lips and, therefore, frequently suffered with drooling of his saliva. He had been prescribed hyoscyne patches to help with the management of this.

During Frank's assessments, he expressed his wish to gain weight and achieve 70kg - his self-reported usual weight. This goal was accounted for in his subsequent care plan.

### Dietetic Management

To help Frank achieve an adequate nutritional intake, food fortification, nourishing snacks and homemade milkshakes were recommended. However, this was not sufficient to make up his significant nutritional deficit. Therefore, an oral nutritional supplement (ONS) was prescribed to prevent further weight loss. Calogen was indicated at 3x30ml per day (405kcal, 90ml fluid) to help him maintain his weight at 64kg.

However, due to his declining condition, Frank was likely to face further challenges with his swallow and chorea movements impacting his nutritional intake and leaving him at nutritional risk. Therefore, enteral tube feeding was discussed with Frank and his parents. The multidisciplinary team (MDT) – comprised of the dietitian, a speech and language therapist, a consultant neurologist, and a regional Huntington's disease support advisor – agreed that Frank was a suitable candidate for gastrostomy placement. Further education regarding the PEG insertion and after-care were undertaken by the dietitian with Frank, his family, and nurses at the care unit. A best care decision was reached and a

percutaneous endoscopic gastrostomy (PEG) was placed soon after.

### Enteral Tube Feeding

Following the insertion of the PEG the goal of dietetic management was to reach and maintain a body weight of 70kg whilst maximising Frank's quality of life. It was important to Frank that he maintained the ability to continue his regular activities, including watching the Formula 1, visiting family, going out to the garden, socialising with his friends at the Neurology Unit, and going on holidays abroad. His feeding regimen was developed, with careful consideration of timing and duration, to have minimal impact on his lifestyle.

The PEG was to be used for supplementary fluids daily and for nutrition when required, to help maintain Frank's enjoyment of food and quality of life.

The regimen was designed to provide a 5x120ml water flush (600ml fluid) each day. Frank's oral intake varied, and when it left a deficit, Nutrison Concentrated was given:

- If less than half of his meals were eaten, a 200ml bolus of Nutrison Concentrated (400kcal, 15g protein) was given.
- If three quarters of the meal was eaten, a 100ml bolus of Nutrison Concentrated (200kcal, 7.5g protein) was given.

Frank continued to take 3x30ml Calogen (405kcal, 90ml fluid) per day orally.

## Ongoing Management

After following his care plan, Frank's weight increased to 68.2kg with a BMI of 19.9kg/m<sup>2</sup>. However, Frank's oral intake declined further (estimated 180kcal per day) and his chorea movements became worse during the night. Overnight feeding increased the risk of aspiration and accidental removal of the PEG, so the feed was increased and restricted to daytime only.

Frank's regimen was altered to 1200ml of Nutrison Concentrated (200kcal, 90g protein) at 200ml/hour over 6 hours during the morning in addition to small tasters of smooth puree texture food orally during the day. Frank would be in his wheelchair and arrangements were made for the feed and pump to be contained within a Flocare Infinity Go Bag attached to the back of his wheelchair. Calogen was given via the PEG instead of orally. This regimen provided a total of 2805kcal, 90g protein, and 1290g fluid per day.

The following year, Frank's chorea movements increased in severity leading to recurrent PEG displacement, therefore his PEG was replaced with a low profile gastrostomy to help reduce the risk of accidental gastrostomy displacement. This way it could be easily reinserted by a nurse should displacement occur. At this stage Frank weighed 69kg and, in an attempt to reach his target weight, his feed was increased to 1500ml of Nutrison Concentrated as well as the 3x30ml Calogen (3405kcal, 113g protein, 1590ml fluid) and additional oral food tasters, dependant on his swallow function. His oral intake also included 1 pint of Guinness throughout the day for pleasure. This PEG feeding regimen allowed him to gain further weight and achieve 71.4kg.

## Outcomes

With the use of nutritional support and a range of medications to manage his Huntington's disease, Frank exceeded his target and reached his peak weight of 75kg and gained better control of his chorea movements. He managed for several years with slight adjustments to his regimen as required.

Over time, Frank's condition deteriorated and in the last few

weeks of his life, his symptoms were monitored and feed volumes waned down to make him more comfortable. His family were very grateful for all the dietetic support he had received over the past 6 years. Frank passed away from his progressive disease at the age of 41.

## Discussion

Accurately estimating the nutritional requirements and designing a suitable enteral feeding regimen for patients with Huntington's disease can be challenging, especially when the patient experiences significant and/or fluctuating chorea movements. Their energy requirements can be high depending on the extent of their involuntary movements and each patient can be different in their display of symptoms and their needs. Increased energy expenditure in Huntington's disease is thought to be due to the following influencing factors:

- An increased BMR and energy expenditure possibly associated with hypothalamic dysfunction<sup>1,2</sup>
- Potential peripheral abnormalities due to the expression of the faulty gene within tissues<sup>3</sup>
- Hyperkinetic movement disorder<sup>4</sup>

There are currently no existing guidelines for estimating nutritional requirements for Huntington's disease patients and further studies within this field are therefore indicated.

Current dietetic practice is guided by close monitoring of weight and body mass index (BMI), changes in the patient's nutritional intake and status together with changes to their clinical presentation. Chorea movements and subsequent metabolic consequences are likely to change and resulting weight loss can be rapid. In addition, there are factors relating to Huntington's disease, such as cognition, mood, swallow function, and ability to self-feed that can have a significant reducing effect on appetite and nutritional intake. Consequently, high energy tube feeds, such as Nutrison Concentrated, are often indicated.

As a result, weight-loss has been commonly reported within the Huntington's disease patient population, particularly in later stages of the disease. Research has shown

the prevalence of these difficulties in this patient group; a study of 63 Huntington's disease patients concluded that 68% of patients experienced weight loss<sup>5</sup>. Dietetic intervention needs to reflect the nutritional goal of the patient, which may be to gain or maintain weight or minimise weight loss. The study also found 25% of patients experience dysphagia<sup>5</sup>. In view of this, and the likelihood of significant nutritional deficit, enteral tube feeding may be required to preserve nutritional status, particularly as the disease and its symptoms progress.

## Conclusion

Frank's case highlights the difficulties faced providing adequate nutrition in Huntington's disease patients. Due to the complexities of this disease, all management options must be explored as different feeding regimens may drastically impact the patient's weight fluctuations and subsequent adjustments of their feeding regimens is essential to provide sufficient nutrition and maintain quality of life. Although Huntington's is progressive and difficult to manage, this case study highlights the importance of patient centred, multidisciplinary care to support these patients through their disease journey and indeed during the final stages of their life.

\*Fictitious name.

## References

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**Nutrison Concentrated is a Food for Special Medical Purposes for the dietary management of disease related malnutrition in patients with fluid restrictions and must be used under medical supervision.**