Huntington Disease

Principles and practice of nutritional management

By Jiří Klempíř and Alžbeta Mühlbäck

Coauthors: Eva Baborová, Petra Havrlíková, Martina Dvořáková, Lucie Růžičková, Romana Konvalinková, Olga Klempířová

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Authors (c):	Jiří Klempíř, Alžbeta Mühlbäck
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Abbreviations

BMI	Body Mass Index
Chap.	Chapter
EHA	European Huntington Association
EHDN	European Huntington's disease Network
e.g.	lat. exempli gratia, "for example"
Fig.	Figure
FEES	Fiberoptic endoscopic evaluation of swallowing
HD	Huntington's disease
NGT	Nasogastric Tube
PEG	Percutaneous Endoscopic Gastrostomy Tube
RMT	Respiratory Muscle Training
Tab.	Table
VFSE	Video fluoroscopic swallowing examination

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Foreword

Food and eating is to all of us an invaluable part of quality of life. In all known cultures meals and food represents pleasure, joy and celebration. Eating together is an important social glue. It is hard to imagine life without any pleasure related to food, but for people affected by HD, meals and food can be experienced as a struggle and cause great stress and despair.

From my own experience I witnessed the struggle my mother had as her disease progressed more than 20 years ago and how today, understanding and knowledge makes my sisters everyday life and eating so much easier. In her late stage of HD she gets appropriate food and help to manage the meals and she still enjoys eating because her carers knows how to prepare her food and assist her in eating.

This book gives patients and the carers loads of advice to how you can adapt your eating and meals to the changes of needs you experience as HD progresses. Managing symptom alterations and adapt to them is crucial help to patients in order to maintain the best possible health condition for as long as possible - not to mention best quality of life.

I recommend the book to everybody. It is well structured and written in an understandable language for all, being a patient, a family member or professional.

Astri Arnesen
- President European Huntington Association -

Good clinical practice is all about making insights productive and impactful in daily life. Huntington Disease presents many challenges to affected and caregivers alike and requires a team of professionals working together to appropriately address the many facets of HD.

It is therefore of great practical impact to compile our current understanding and the practical advice of experienced health care professionals in a well-structured fashion and in an easily accessible language thus facilitating communication between patients, family members and professionals. Learning to adapt to changing symptoms and signs is critical for maintaining quality of life and participating in life throughout the course of HD.

I would therefore like to wish the book by Jiří Klempíř and Alžběta Mühlbäck a good reception, a wide circulation and a long lifecycle with regular updates reflecting the continuing progress of further improving care for HD.

Prof. G. Bernhard Landwehrmeyer, MD, FRCP
- HD Center Ulm, Germany, Principal Investigator Enroll-HD –

1. Introduction

by Jiří Klempíř, Alžbeta Mühlbäck

Huntington's Disease (HD) is a neurodegenerative disease that manifests in changes of movement disorders and mental functions. Some of the symptoms of the disease can be alleviated, but there is still no cure available to stop the progression of HD.

Huntington's Disease does not affect only the nervous system, but also other tissues in the body in different stages. All patients will, in the course of HD's progression, experience problems with proper nourishment and weight loss.

Every organism needs to receive and process energy and nutrients in sufficient quantities to maintain its life functions. All chronic diseases, including HD, lead to increased demand for energy and nutrients. If these needs are not continuously and adequately met, the organism starts to suffer from **malnutrition**. In such cases, patients are tired and weak, prone to infections and other medical complications; they cannot sufficiently participate in supportive physical therapies or benefit from any treatment compared to patients without nutrition problems. When malnutrition is detected early and proper steps are taken, it can significantly improve the patient's quality of life.

The aim of this publication is to summarize all relevant and practical information to enable the early detection of nutritional disorders as well as their causes and consequences. Additionally, it provides practical recommendations for how to proceed in such cases.

The publication is in form and content primarily addressed to patients, their families and caregivers or medical staff to receive information on the issue of nutritional disorders within Huntington's Disease.

2. What is Huntington's Disease?

by Jiří Klempíř, Olga Klempířová

Huntington's Disease is an inherited disease characterized by progressive movement disorders, the decline of intellectual abilities and behavioral changes. Among the Caucasian population it is estimated that the mutation prevalence is about 1:15,000.

2.1. What causes Huntington's Disease?

Huntington's Disease is caused by gene mutations on chromosome 4. The gene product is a protein called Huntingtin which occurs naturally in all cells. Huntingtin is needed for proper development of an organism. Mutated Huntingtin has different characteristics, and its products are considered to be toxic. The presence of the mutated gene does not automatically cause illness, however, it depends on the severity of the mutation. The mutation carriers usually live in good health until the onset of the disease.

The age of the clinical onset of the disease depends on the properties of the mutated gene and other genetic factors of the individual. In rare cases, Huntington's Disease may begin during childhood or late into old age, but most typically its onset is in young adults and middle aged adults. The offspring of mutation carriers are 50% likely to inherit the mutated gene. The presence of the mutation can be reliably determined by a genetic test with a blood sample.

2.2. Movement disturbances

HD, in the same way as other neurodegenerative diseases, develops symptoms slowly over a period of many years before showing the first, unmistakable signs of the disease. Huntington's Disease is characterized by the presence of **choreatic involuntary movements** (greek: chorea = dance) that occur as irregular, jerky movements in different parts of the body. Their intensity and severity may vary from discrete jerks in the fingers up to excessive large-scale movements affecting stable posture and gait and disturbing daily activities. With the progression of the disease, the involuntary movements lessen and become slower, and the **slowness of the voluntary movements** becomes more significant (bradykinesia) together with an **increase in muscle tension** (rigidity). In advanced stages of the disease, the patient is very limited in his or her mobility and is usually bedridden.

2.3. Cognitive disorders

Patients with Huntington´s Disease's also experience a cognitive decline. Cognitive changes may occur years before the onset of visible movement symptoms. Among the first mental changes is the **failure to recognize emotions**, especially negative ones (fear, sadness, disgust). Almost simultaneously, difficulties in recognizing speech intonation and its meaning, and nonverbal communication (ie. body language) increase. All this together results in conflicts and misunderstandings in patients' work and private life.

In subsequent years, the following disturbances start to occur: reduction in **attention**, problems in **perception of time** and **planning**, and generally the **speed and flexibility of thinking** in new or challenging situations, slows down. Patients and their relatives become aware of problems with **learning new information** and physically demanding tasks. Some patients, however, **deny, trivialize or do not perceive their behavior and physical health** (anosognosia). Deepening cognitive deficits gradually increase the patient´s dependence on his or her caregiver.

A large problem with patients with HD is apathy. **Apathy** is the loss of interest in oneself and surroundings, the inability to actively start an activity and successfully complete it. This is not fatigue, with which it is often confused. Sometimes apathy contributes to depressive feelings. Apathy often becomes a major obstacle for motivation to do various activities, including employment, exercise, leisure activities and also the patient's self-care.

2.4. Behavioural disorders

Throughout the course of the disease, changes in personality and a variety of emotional and behavioral disorders may be present. Very common are **anxiety** and **depression**, **impulse control disturbances**, verbally or physically **aggressive behavior**, **psychotic symptoms**, alcohol and nicotine **abuse**, and rarely, minor criminal offenses.

Behavioral disorders and cognitive decline often cause more serious problems than the change of movements for patients and caregivers.

2.5. Disease´s Variance

The course of the disease will vary according to the age of the disease's onset.

An **adult onset of Huntington's Disease** affects 90% of all patients and is described in the previous text.

Children and adolescents are affected by **Juvenile Huntington's Disease** (Westphal variant). It is characterized by significant slowness of movements (bradykinesia),

increased muscle tension (rigidity), minimal presence of chorea, rapid cognitive deterioration as well as aggression and other behavioural changes similar to Schizophrenic disorders (hallucinations, delusions).

The **senile form** (late onset) of Huntington's Disease begins after the 65th year of age with usually a mild choreatic syndrome and moderate impairments in cognitive and behavioural functions. Due to the late onset of disease and its very slow progression, patients are self-sufficient for many years and usually do not die from complications associated with Huntington's Disease.

2.6. Malnutrition and its complications

After years of enduring the disease, patients experience chewing and swallowing disorders with increased energy turnover leading to malnourishment. These changes cause an overall weakening of the body, including respiratory muscles and the immune system. Dysphagia (swallowing problems) often lead to leakage of fluids and food into the windpipe (**aspiration**). Patients are at risk of choking and developing inflammation in the lower windpipe.

Loss of muscle mass and fat deposition together with immobilization lead to the formation of bed sores (**decubitus**). Local infections (e.g. from wounds, the urinary tract) can easily get into the blood stream and cause sepsis and a total failure of the body, since it is too weak to handle these complications.

Summary

- The mutation carrier of HD is born healthy and develops normally
- Each direct descendant of a mutation carrier has a 50% risk of becoming a carrier of the same mutation
- Huntington's Disease manifests in movement, behavioural and cognitive disorders
- Patients experience problems with their liquid and food intake and experience weight loss as a result. Early detection can help to prevent resulting complications.
- Ensuring safe and adequate intake of energy and nutrients improves the quality of life

3. Basics about nutrition

by Jiří Klempíř

3.1. The energy requirements of the human body

Every organism needs a minimum caloric intake, called basal energy expenditure, to carry out the basic metabolic functions. **Basal energy expenditure** for a male is 25-30 kcal/kg/day and for women about 10% less (1 calorie = 4.185 joules). In practice, this means that a person weighing 70 kg should consume 1500-2000 kcal per day. Furthermore, it is necessary to take the supply of **energy for physical and mental activity** into account. There is an energy requirement for processing food intake (thermal effect of food). A healthy and active person usually consumes 2200 – 2600 kcal per day.

The total energy turnover is strictly individual and influenced by many factors such as: genetic makeup, hormones, diet, lifestyle and environmental factors. Under certain circumstances, such as increased physical activity, mental stress or illness, the energy consumption may increase up to two or three times. Generally prolonged energy excess leads to weight gain and restricted energy to weight loss.

3.2. Importance of basic components of nutrition

The basis of healthy nutrition is a balanced intake of **macronutrients** (proteins, carbohydrates, fats) and **micronutrients** (minerals, vitamins, dietary minerals). Their consumption may vary individually according to age, physical or psychological stress, pregnancy, etc. There are many high-quality publications available on the topic of healthy nutrition, hence here we will focus only on the most important facts.

3.2.1. Carbohydrates

Carbohydrates (sugars) serve as the main energy source for the body (1 g carbohydrate = 4 kcal = 16.8 kJ) and cover between 40 - 60% (approximately 4 g / kg / day) of the energy requirements. They can be divided into simple and complex carbohydrates.

Simple carbohydrates include glucose and fructose (fruit sugar in fruit and honey) and sucrose (white sugar). Simple sugars are rapidly absorbed into the blood; they lead to an increased insulin secretion and subsequently sharp decline in glycaemia, inducing the feeling of hunger.

It is better to consume **complex carbohydrates** (polysaccharides) that are absorbed slower and hence the feeling of satiety lasts longer. Polysaccharides manifest in the form of starches in bread, rice, potatoes, pasta as well as in vegetables.

3.2.2. Lipids

Lipids (fats) cover 25 to 35% of energy intake (approximately 1 g / kg / day) and provide a high-energy content (1 g = 9 kcal = 37.8 kJ) which is processed slower in comparison to sugars. Lipids are by origin divided into plant and animal lipids.

Most **vegetable fats and oils** are more suitable for nutrition due to a higher content of unsaturated fatty acids. **Unsaturated fatty acids**, especially omega-6 and omega-3 fatty acids occurring in certain plants and particularly in fish oils, help to reduce cholesterol levels. Omega-3 fatty acids reduce the triglyceride concentration in the blood plasma and improve certain other cardiovascular parameters.

Animal fats (lard, butter) are a source of trans-fatty acids. An increased intake of animal fats can cause higher cholesterol levels. Cholesterol is as well important for the construction of cellular membranes, the nervous system, the formation of some hormones and also vitamin D. During growth or regeneration, its consumption in the body rises.

3.2.3. Proteins

Proteins supply the smallest amount of energy, around 12 to 15% (approximately 0.8 g / kg / day), with an energy value similar to that of carbohydrates (1 g = 4 kcal = 16.8 kJ), and they are necessary for the development, growth and regeneration of the organism. Proteins are parts of enzymes and hormones, and are involved in the transport mechanism of different substances in the body.

Animal proteins, unlike vegetable proteins, are considered to be perfect as a source of all known essential amino acids that our body cannot produce and need to be obtained through a balanced diet. The ratio between animal and vegetable proteins should be optimally 1:2, and in the case of an increased protein consumption in the organism, 1:1 is recommended. Under ongoing or prolonged stress conditions (serious illness, convalescence) the organism´s protein consumption is increased to 1.6 to 2.2 g / kg / day.

3.2.4. Vitamins, minerals and dietary minerals

Micronutrients such as **vitamins**, **minerals** and **dietary minerals** form an integral part of a healthy diet and with only a few exceptions, the human organism cannot produce them. Micronutrients are essential for the proper function of various cellular processes including immunity and antioxidant mechanisms.

If a healthy person consumes a balanced and varied diet including fruits and vegetables, and is not exposed to extreme stress and conditions, they will not suffer from a serious lack of micronutrients. However, an excessive ongoing intake of micronutrients in the form of multivitamin mixtures may have adverse effects on the

body (e.g. digestive problems, increased oxidative stress - "oxidative paradox"). It is recommended that the content of micronutrients in a single daily dose should not exceed more than half of the recommended daily allowance.

Table salt contains sodium, which is necessary to maintain the water balance in the body, but its excessive consumption can cause serious health problems and burdens the cardiovascular system and the kidneys. Salt, for its flavour and preservative properties, is added to most dishes. In the Western world, the average consumption is around 12 to 20 grams daily, although the recommended daily dose for adults is only 5-7 grams. One teaspoon contains about 8 grams of salt.

3.2.5. Dietary fibre

Fibre plays a special role in the human diet. Edible parts of plants are resistant to digestion and absorption in the stomach and the small intestine of the human body, but completely or partially decompose in the colon.

Dietary fibre consist of a mixture of non-starch polysaccharides and many other plant constituents such as cellulose, lignin, chitin, pectin, beta-glucans and oligosaccharides.

Soluble fibre (soft, digestible) have the ability to absorb water (pectin, inulin). The short chain fatty acids are produced by bacterial fermentation of dietary fibre. The short-chain fatty acids promote a suitable environment for acidophilic bacteria. Therefore, these types of fibre are known as **prebiotics**.

Insoluble fibre does not dissolve in water, acts as a cleansing agent, improves the bulk and consistency of stool, shortens intestinal transit time (relieves constipation) and reduces the incidence of dental decay (cellulose, lignin). Fibre binds to toxic substances and decreases the absorption of fats and cholesterol as well. Therefore, fibre is recommended for the prevention of cardiovascular diseases, tumours of the digestive tract and diabetes.

The recommended daily dose of fibre is 30-35 grams/ day and is covered with a consumption of 500 grams vegetables and fruits.

3.2.6. Probiotics

Probiotics are live physiological bacteria and yeast. They help to restore the balance of microflora in the digestive tract. Lactobacillus, Bifidobacterium, Escherichia coli and some of Saccharomycetes, Streptococcus, Lactococcus, Enterococcus are all probiotic bacteria. A similar effect to that of natural probiotics can be achieved by the regular use of probiotic products. Probiotics are administered in case the gut microflora are disturbed or in danger of an overgrowth of pathogenic bacteria, e.g. after treatment with antibiotics or an inflammatory bowel disease.

Summary

- On average an adult consumes 2200 – 2600 kcal per day
- The human organism needs a proper mixture of plant and vegetable proteins, fats and sugars in the right quantity and form
- The diet should be varied, balanced and tasty
- A balanced diet includes plenty of fibre, minerals, vitamins and dietary minerals
- Physical and mental stress and illness increase energy and nutrient consumption
- Under ongoing stress, energy consumption also increases.

4. Hydration

by Jiří Klempíř

4.1. The importance of water for the human organism

The proportion of water in an adult body is 60 to 70%, in early childhood even up to 80%. Water plays an important role in dissolving substances, cell metabolism and the transport of nutrients and as well in the elimination of waste and harmful substances from the body. Naturally the human body loses water through breathing, sweating and excretion of urine and feces.

To maintain the proper amount of fluids in the body, it is necessary to keep drinking an adequate amount of water and have a proper water regime, which varies according to age and body burdens. For adults, the **recommended water consumption is usually 2-3 liters per day**, including liquids from food and water produced by processing of substances in the body (about 250ml / 24 hours).

4.2. Causes and signs of dehydration

Dehydration (water deficit in the body) occurs, when the organism loses more fluid than it receives. During dehydration, the body does not have enough fluid to function normally.

Dehydration usually develops over a few days. An adult survives without fluid intake for 2-4 days. Children can develop dehydration even within a few hours. Dehydration can also be caused by organic diseases such as kidney disorders or high blood sugar levels over a longer period of time.

Reduced intake of fluids in some instances may be caused by a lack of proper fluid intake for hydration of the organism or due to consumption of food and dietary products with a high amount of salt, minerals and sugars. Usually patients having problems with urinary incontinence tend to have a reduced intake of fluids. In the case of Huntington's disease, the main causes of inadequate hydration are: Apathy, with no interest in daily activities, and memory and behaviour problems. Sometimes swallowing problems are hidden causes of dehydration.

Increased loss of fluids often arises from excessive consumption of caffeine (coffee, strong tea, coke and other caffeine drinks) or by the use of diuretic medication for the treatment of high blood pressure or renal dysfunction. Acute dehydration may also occur suddenly in cases of vomiting, diarrhoea, sweating from fever, and high temperature of the environment.

The spectrum of symptoms of dehydration may vary individually according to age, the original condition of the person, associated diseases and depends on the level of dehydration (Table 1, 2).

Table 1: Symptoms of mild dehydration

Intense thirst
Dry mouth
Concentration problems, fatigue
Tiredness
Headache
Dizziness
Concentrated urine and less in quantity
Pre-collapse conditions

Table 2: Symptoms of severe dehydration

Intense thirst
Extreme tiredness, lethargy, weakness
Irritability
Disorientation / confusion
Absence of sweating
Reduced skin tension
Increased body temperature
Decreased urine volume
Increased heart rate
Low blood pressure Collapse conditions
Circulatory failure and death

4.3. Therapy for dehydration

Mild dehydration (deficit from 0.5 to 2.0 l) recovers after administration of an adequate fluid quantity and, consequently, the general condition improves on the same day.

Severe dehydration (fluid deficit over 3.0 l) can be a very serious condition even for a healthy person and requires urgent hospital care. The patient undergoes a comprehensive examination and after evaluation, controlled rehydration will begin. In critical condition, it may take several days. Rapid hydration could have fatal consequences.

4.4. Prevention of dehydration

It is important to take fluids in small doses throughout the whole day. Surge intake of large amounts of fluids can cause nausea. On a trip outside home, it is important to carry a bottle of water along. During stress periods or when ill, it is necessary to increase the intake of appropriate fluids. In the case that a person has problems remembering or is unsure about his or her own fluid intake, it is recommended to write down the amount of all fluids consumed during the day using a journal. In the evening, an evaluation can be done. To have a clear impression of fluid intake, the journal should be written over a longer period of time. Another option is to prepare the daily dose of fluids to be taken in advance to achieve proper hydration in the morning (e.g. as ready-made water bottles).

It is desirable that even people who have problems with self-care, should have an easily accessible option to have a drink anytime. For these cases, small bottles with a teat or a straw, the same as used for infants and toddlers, are suitable. Swallowing problems can be improved through a straw when drinking, and chilling drinks or modifying their consistency through the use of thickeners can also help (see chapter 6).

If the person regularly receives a high quality and balanced diet, then he normally does not suffer from a lack of minerals that help maintain the physiological distribution of fluids in the body. Fruits and vegetables contain 70-90% water as well as extra fibre, vitamins and minerals. During extreme bodily stress (athletic performance, overheating, vomiting, diarrhoea), there can be significant losses of minerals, and in such cases, it is not appropriate to drink only pure water but also fluids containing a higher mineral amount.

The measure of appropriate hydration is the colour of urine. If the urine is a clear, light colour hydration is sufficient. If urine is dark in colour and not much quantity, the patient's drinking regime should be increased. If the patient has no impairment of renal function or heart failure, an increased fluid intake is considered to be safe.

Summary

- The recommended daily fluid intake for an adult is 2 – 3 litres
- One should drink slowly in small doses (100 - 300 ml) throughout the day
- Vomiting, diarrhoea, increased physical activity, fever and high temperature of the environment increase the demand for fluids and minerals
- Mild dehydration (deficit 0.5 - 2.0 l) normally gradually improve on the same day
- Severe dehydration requires urgent hospital care

5. Malnutrition

by Jiří Klempíř

5.1 Definition and occurrence of malnutrition

The term **malnutrition** means poor nutrition and concerns people that are malnourished, obese and with inadequate intake of certain food components.

In general, malnutrition arises from various causes and is not a rare phenomenon even in civilized countries. Children, the elderly and patients with occurrence of one or more chronic diseases have an increased risk for malnutrition. Approximately one third of patients who are admitted to hospital for various reasons suffer from malnutrition. Among long-term hospitalized patients or among nursing homes patients, malnutrition occurs in half of the patients.

Obesity is rarely encountered in Huntington's disease cases and usually does not have serious consequences. The following text is therefore focused mainly on an insufficient intake of energy and nutrients. Malnutrition sooner or later affects all people with Huntington's disease. Effective prevention, early detection of malnutrition, and its proper treatment improve the quality of life for patients.

5.2. Causes of malnutrition

Malnutrition may arise for various reasons. Mostly it is caused by reduced food intake or increased consumption of energy and nutrients (Table 3).

Table 3: Causes of malnutrition

Inadequate food intake due to: Insufficient quantity and quality of balanced diet, poor condition of the teeth status, allergies, swallowing disorders, lack of appetite, side effects of treatment, psychiatric illness, dementia, low economic status

Digestion problems: Lactose intolerance, allergies, inflammation of the intestines or pancreas

Metabolic disorders: Diabetes, liver or kidney failure, thyroid disorders

Increased loss or consumption of nutrients and energy: Fistulae, abscesses, infection, trauma, surgery, cancer

5.3. Types of malnutrition

There are two basic types of malnutrition, which may have various causes.

Simple malnutrition (undernutrition/starvation) develops gradually as a simple adaptation of the organism to **decreased intake and simultaneously reduced energy expenditure**. This type develops slowly and gradually during weeks to months and presents with weight loss associated with the reduction of body fat and protein, including muscle mass and fluid retention. Some typical examples are senile cachexia, anorexia nervosa and undernutrition in cancer and neurodegenerative diseases.

Stress malnutrition (stress starvation) occurs by **increased energy consumption and the lack of energy intake**. If there is an acute stress condition, initially the body weight does not have to change, but proteins may be quickly consumed, because carbohydrates are more difficult to process and the body cannot effectively process the fat reserves. Patients in critical condition may lose about 250 grams of their muscle mass daily. However, the protein deficiency does not only manifest as an overall weakness of the limbs muscles, but as well as cardiac and respiratory muscle deficiency. Therefore, stress malnutrition can seriously affect the patients' health status. Among the common causes of stress malnutrition are sepsis, acute abdomen cases and multiple trauma.

5.4. Specific causes of malnutrition in Huntington's disease

Various factors are involved in the development of malnutrition in Huntington's Disease and some of them are very typical for this disease.

The basal energy expenditure progresses already several years before an onset of the Huntington's Disease (see chapter 3.1). Despite a normal food intake, the patients may achieve a negative energy balance leading to unwanted weight loss.

Although Involuntary movements or increased muscle tone (rigidity) seem to consume more energy, they have no significant effect on total energy expenditure. However, even if involuntary hyperkinetic movements are increased, the patient´s total motor activity decreases due to apathy and lack of interest. Voluntary movements slow with the progression of the disease (bradykinesia) and the patient requires more effort to perform these movements, even for daily activities. Bradykinesia, which is often covered by involuntary movements, continuously increases together with weight loss.

Development of malnutrition can negatively reinforce the widely occurring **swallowing disorders** (see chapter 6). The swallowing act may be disrupted by involuntary movements of the lower jaw, tongue (restless tongue), pharynx, larynx and respiratory muscles. Involuntary movements of the mandible can lead to destruction of normal or artificial teeth and to dentures instability. Sometimes hyperphagia (excessive hunger or

increased appetite) can occur and the patient tends to swallow bulky bites or inedible objects. All of these disorders can lead to manifest or silent aspirations, including asphyxia.

Regular food intake may be distracted by significant **cognitive deficits** (see chapter 2.3), **behaviour disorders** (see chapter 2.4), **financial problems**, **poor family background**, **poor knowledge of the importance of nutrition** and **inadequately educated medical staff**.

Patients with Huntington's Disease, in comparison to healthy individuals, showed a higher incidence of clinically symptomatic and asymptomatic oesophagus and stomach inflammation, and disturbances with gastric emptying, which causes nausea and a reduced appetite.

It is not yet clear to what extent mutant huntingtin influences an increased energy turnover, negative balance, degeneration of certain brain structures associated with eating behaviour and disorder of hormones, cholesterol, lipids and fatty acids. Similarly, as in the case of mitochondria, cell structures are responsible for the production of energy in a living organism.

5.5. Complications of malnutrition in Huntington's Disease

Acute and chronic diseases can cause malnutrition and enhance the already present symptoms of malnutrition. Malnutrition itself may lead to additional complications and significantly reduces the quality of life (Table 4)

Table 4: Complications of malnutrition

Reduces physical resistance
Reduces mental endurance
Causes and exacerbates fatigue and apathy
Prolongs the duration of acute disease
Cause complications in the course of acute and chronic diseases
Prolongs the stay in hospital
Prolonged recovery
Increases mortality

5.6. Diagnosis of malnutrition

Proper and early identification of factors contributing to malnutrition can lead to effective prevention and treatment. Basic tests are either complicated or difficult to perform, they are usually initiated and done by a doctor or a nutrition specialist. The basic examination consists of medical history (anamnesis), and physical and laboratory examinations.

5.6.1. Anamnesis

To obtain an **anamnesis** means to receive detailed information of the medical problem and its relations in a personal interview. It may happen in some cases that the information obtained from the client / patient may be for various reasons incomplete or misleading (shame, denial, communication problems, memory disorders, mental illness, etc.). It may help to talk to family members and caregivers to complete the anamnesis. In terms of nutrition, the most important aspect to focus on is the **detection of unwanted weight loss**, and **its dynamics** (see table 5). At the same time, asking about eating habits, diet, the amount of food and any dietary restrictions (see chapter 7) is important. Patients are often not prepared for these questions and have difficulty answering, then it is necessary to ask what they ate in recent days, the last 24 hours, if they noticed loss of muscle mass, loose clothing, etc. The weight loss is often associated with disorders of the digestive tract, which appear as: loss of appetite, chewing and swallowing problems, heartburn, eructation (belching), hiccups, bloating, pain, and abnormal bowel. It is important to know about any substance abuse (nicotine, alcohol and other drugs). It is as well important to know about past or ongoing diseases, trauma, surgeries, infections, increased temperature without clear causes and medications (chapter 5.2, chapter 12)

Table 5: Risk of malnutrition developing

Insufficient food intake
Missing food intake for more than 7 days
Food intake <70% of the required amount for more than 10 days
It is not possible to receive food in sufficient quantity and quality
Weight loss
Weight loss > 10% over the last 6 months
Weight loss > 5% in the last month
Decrease in body weight > 20% absolute

Lack of physical resources

Any weight loss with BMI <18.5

Loss of muscle mass associated with the loss of self-sufficiency (e.g. bedridden, not leaving home)

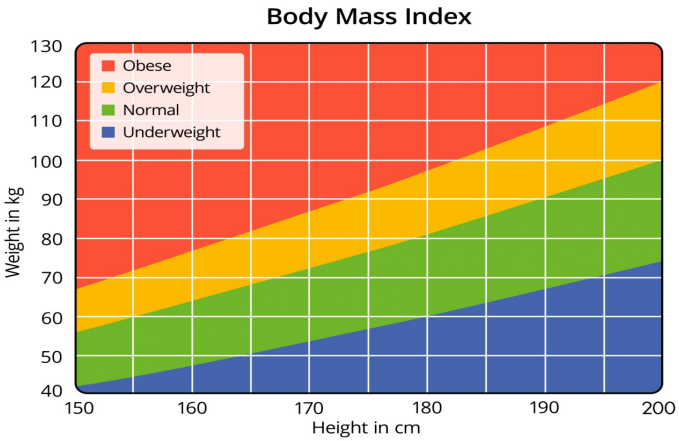
5.6.2. Physical Examinations

A basic **physical examination** serves as evaluation of physical condition. The constitution and the condition of skeletal muscles are evaluated visually. While other symptoms or causes of malnutrition, edema in the lower limbs, retention of free fluid in trunk, skin defects, bruising, inflammation in the oral cavity are verified.

Part of the examination is to calculate the **body mass index** (BMI). The BMI is determined by the following formula:

$$BMI = \frac{\text{weight (kg)}}{\text{height (m) x height (m)}}$$

Figure 1: BMI graph



The BMI provides only a simplified view of the current condition of the body, as it does not consider the body's constitution, such as the muscle mass and adipose tissue. For example, a bodybuilder may, due to a large amount of muscle mass, fall into the category of obese people with a BMI measurement. BMI also provides invalid information for obese persons with a small proportion of muscle.

As people age, they naturally lose muscle mass, strength and function over time. This phenomenon is called **sarcopenia**. As well, obese individuals may be impaired by significant loss of muscle mass. In this case, the condition is called **sarcopenic obesity**, when the total weight may stagnate or even be increasing, through the increase of fat tissue. An organism under distinct stress load (see chapter 5.3) is unable to effectively use the fat tissue, and therefore it is the most dangerous kind of obesity. Prevention of obesity and sarcopenia are adequate diet and physical activity.

The proportion of muscle and adipose tissue and muscle function may only be reliably and noninvasively examined in specialized nutritional and sports labs and clinics.

Cachexia is significant weight loss, usually with a BMI ≤ 18.5 points. A BMI ≤ 16.5 points and indicates severe malnutrition and such an individual, even if otherwise healthy, is in a life-threatening condition. Literature usually states that a BMI 20.0 to 25.0 is considered to be a normal condition, but this rule may not acquire for impaired persons. For example, patients who are overweight should not reduce their BMI below 23.0 and in the case of individuals with a BMI <23.0 it should not drop below 20.5 points.

5.6.3. Laboratory findings

Laboratory tests are initiated by the doctor in order to obtain more detailed information on the nutritional status and to exclude or confirm causes that cannot be found by above procedures.

In practice, the following tests apply: Sedimentation and biochemical parameters of blood, blood count, biochemical and infectious parameters in urine. Depending on the circumstances, a physician may indicate further supplementary examinations (X-ray, ultrasound, endoscopy etc.) or consult a specialist.

In the case of simple starvation (chapter 5.3.), the blood test has only limited importance, and may rarely be used to monitor micronutrients (chapter 3.2.4.). On the other hand, it is important to follow laboratory examinations during starvation stress (chapter 5.3.), because the results help determine the treatment strategy of malnutrition and plans for recovery.

The above process requires no special equipment or training. Therefore, early detection of developing malnutrition and its prevention or early detection of serious

unrecognized diseases (e.g. cancer), may be done by the family doctor or a general practitioner.

Summary

- Weight loss is an integral part of Huntington's Disease
- Malnutrition may also occur for other serious reasons
- Malnutrition weakens the mental, physical and immune resistance of the organism
- Malnutrition worsens the course of illness and may itself cause further complications
- The body weight is the best parameter for early detection of the development and treatment of malnutrition
- In case of illness, it is useful to regularly monitor the body weight
- Optimal BMI is between 23 - 25
- For obese patients, BMI may be misleading
- For the general health, well-being, and quality of life it is important to maintain a sufficient amount of functional muscle mass
- A balanced diet and physical activity is the best prevention of muscle loss
- In case of unwanted weight loss, it is necessary to determine the cause and to prevent the development of malnutrition
- Nutritional screening may be done at home and further be consulted with a physician in order to perform additional tests, if needed
- If one is unable to maintain optimal weight, it is advised to consult a nutrition specialist

6. Dysphagia – swallowing problems

by Eva Baborová

6.1. Introduction

A healthy person swallows about 520 times a day without thinking about it. Swallowing is a complex process that requires coordination of almost all oral and facial muscles with different areas of the nervous system.

Swallowing disorder (**dysphagia**) is a condition describing the difficulty in swallowing. Patients with dysphagia are at risk of serious medical complications.

6.2. Swallowing process

In order to better understand swallowing problems, it is important to understand the swallowing process itself. As noted in the introduction, this is a very complex process of proper muscles coordination and its timing, as it does not take longer than three seconds. The swallowing process is divided into four phases (figure 2):

6.2.1. Oral phase

During the oral phase the food is prepared by chewing and mixing with saliva in the oral cavity and a bolus of smooth consistency is shaped.

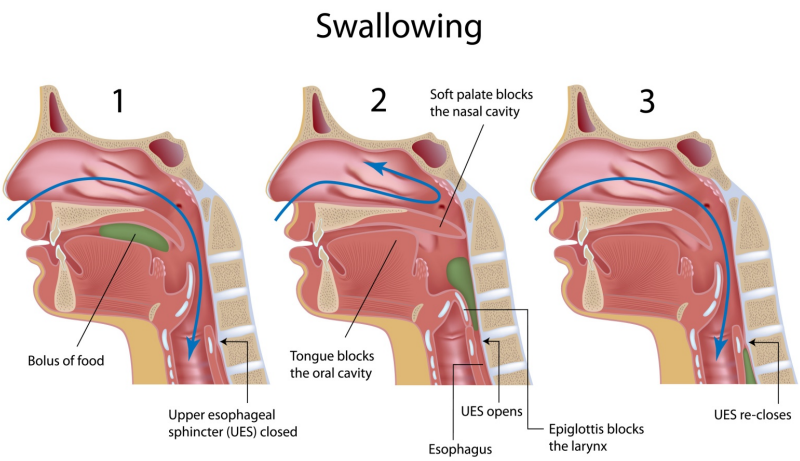
6.2.2. Pharyngeal phase

In this phase the tongue moves the bolus towards the back of the mouth and pharynx. At the same time the soft palate raises to close the nose entrance and the vocal folds (epiglottis) close to protect airways from food and liquid entries. The person stops breathing for a very short time and the bolus moves to the lower part of the pharynx.

6.2.3. Esophageal phase

The upper esophageal sphincter opens and the bolus passes in the esophagus and moves by esophageal muscles contractions into the stomach.

Figure 2: Swallowing phases



6.3. Symptoms of a swallowing disorder (dysphagia)

Difficulties may occur in all swallowing phases. A careful diagnosis is very important to identify and localize the problem.

The most common difficulties occurring in the oral phase include:

- Difficulties with tongue control and chewing of solid food
- Reduced tongue motor abilities (may cause problems with swallowing tablets)
- Problems to create a bolus and move the bolus towards the back of the mouth
- Reduced tongue root motility resulting in stacking of bolus in the throat
- Inability or delayed ability to trigger the swallowing reflex

In the pharyngeal phase: Due to poor coordination and reduced tongue motility the food rests cannot move forward in the throat (stacking) and may enter the airways, causing silent aspirations. The major signs indicating emerging dysphagia are summarized in table 1.

Table 6: Clinical signs of dysphagia

Coughing
Chocking
Elevated temperature
Weight loss
Changes in voice quality after swallowing (table 6)
Breath shortness
Nasal regurgitation
Leakage of fluid or saliva from the mouth
Chewing difficulties
Prolonging of meal time

6.4. Aspiration

One of the most serious consequences of dysphagia is **aspiration, when food or fluids from the stomach or mouth is entering the lungs** due to swallowing problems. Each of the following symptoms (table 7) should prompt an immediate medical consultation to determine the presence of serious dysphagia with aspiration.

Table 7: Aspiration symptoms

Significantly increased salivation
Wheezing and hoarse voice
Sudden coughing, "cleaning the substance from lungs''
Increased body temperature
Tearing eyes
Cough with phlegm

Apart from aspiration, we also distinguish the **silent aspiration**, when food, fluid, saliva or stomach contents enter into the lungs and the patient shows no signs of aspiration such as coughing, choking and turning red. If a silent aspiration occurs, the patient is

significantly at risk of lung pneumonia. This condition mainly occurs due to a weakness in the cough reflex, and generally during the progression of Huntington's Disease.

6.5. Swallowing disorder in Huntington´s Disease

During the course of progression of Huntington's disease, voluntary movements and difficulties in their control and coordination gradually worsen, causing swallowing problems. In the advanced stages, the swallowing difficulties can be very serious and require some special management. Patients may experience chewing and swallowing problems, not just with solid foods, but also with swallowing of saliva and other fluids. The patient is thus at risk of suffocation and aspiration pneumonia.

Aspiration pneumonia occurs when foreign particles, in this case, food, fluids or stomach content enter into the lungs. Repeated aspiration pneumonia is a common cause of death in Huntington's patients. Huntington´s disease affects every single phase of the swallowing process. The whole swallowing process is also altered by other symptoms, such as changes in eating habits, memory and concentration problems.

Problems with swallowing cause not only physical complications, but also affect the patient's mental status. The fear of not being able to swallow, resulting in anxiety of not being able to eat and drink normally can overwhelm the patient. This can be avoided by learning the necessary preventive techniques and strategies for better control in advanced stages prior to disease progression.

6.6. Dysphagie examination

For better prevention and management of swallowing difficulties it is important to consult a speechlanguage pathologist for diagnosis and therapy recommendations. The speech-language pathologist examines the patient and identifies problems. They may propose the individual management of swallowing disorders or discuss with the patient and the caregivers the introduction of enteral nutrition. Sometimes imaging examinations such as a fiberoptic endoscopic evaluation of swallowing (FEES) or video fluoroscopic swallowing examination (VFSE) are useful.

6.6.1. Imaging methods

Imaging methods are objective methods for the evaluation of swallowing disorders. These methods can accurately assess all phases of swallowing and target therapies to the particular difficulties.

6.6.1.1. Video fluoroscopic swallowing examination (VFSE)

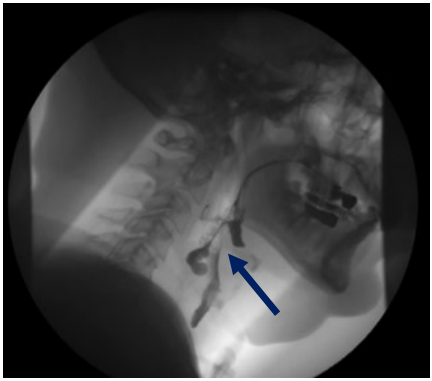
This is a real time X-ray examination that can show the entire course of the swallowing process (figure 3). During this examination, the patient is standing or seated in front of

the X-ray device (fluoroscope) and camera that displays the course of the swallowing process onto a monitor. The patient is given three different food and liquid consistencies with contrasting material, such as: fluid, purée and solid food, that simulate the consistency of a regular diet.

This examination evaluates swallowing difficulties in detail, helps to set the therapy, and identifies the consistencies that patients can most safely consume.

Figure 3: Video fluoroscopic swallowing

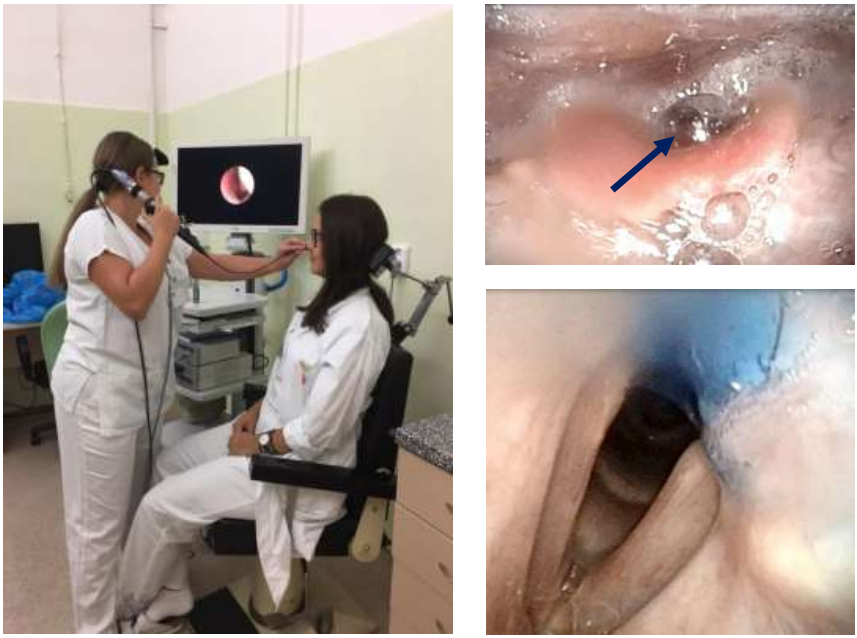
Examination (VFSE) with a form of real-time x-ray imaging (fluoroscopy) showing aspiration (blue arrow)



6.6.1.2. Fiberoptic endoscopic evaluation of swallowing (FEES)

The second method of objective examination is the fiberoptic endoscopic evaluation of swallowing (FEES) or examination with a flexible endoscope (figure 4). This is an examination mostly performed by otolaryngologists or speech-language pathologists. An endoscope is a device that looks like a flexible tube with a light and camera. This camera is inserted through the patient´s nose into the throat (hypopharynx). Again, several food and liquid consistencies are swallowed. The examination is well tolerated; though sometimes light scratching in the throat may occur.

Figure 4: Fiber optic endoscopic
Evaluation of swallowing (FEES) showing a silent saliva aspiration (blue arrow) and an aspiration with the blue-dyed food



6.6.2. Results of the examination

The speech-language pathologist estimates after the evaluation of the examination the results and appropriate strategies for the management of the swallowing disorder, such as:

Effortful swallow:

Collect all the saliva in your mouth in the middle of your tongue. Keep your lips tightly closed, imagine that you have to swallow something big in your mouth (ping-pong ball, large grape) and swallow. The number of repetitive exercises is individual for each patient.

Masako manoeuvre:

Stick your tongue between your front teeth and gently bite down to hold it in one place and try to swallow, while the tongue stays on the same place between your teeth. The number of repetitive exercises is individual for each patient.

Besides regular exercises under the guidance of a speech-language pathologist, the following recommendations are to be followed:

Patient's position when eating and drinking

- Sit up as straight as possible
- Tilt your head slightly forward
- Sit upright for at least 15-20 minutes after eating

Dining environment

- Minimize everything that could distract when eating
- Focus only on food and drink
- Do not talk with food in the mouth

Swallowing

- Eat slowly
- Cut the food into small pieces and chew thoroughly
- Swallow each bite 2 - 3 times
- If the food stacks in your throat, cough shortly and try to swallow it again before you take another breath, repeat if necessary

Food and consistency

- Select soft and smooth food, when chewing is difficult or tiring
- Mash or blend foods, if needed
- Try thickening of liquids, if normal fluids cause coughing after swallowing
- Avoid food of two and more different consistency such as soup or salad with dressing or adhesive food (plain mash potatoes)

Salivation

- Drink enough of fluids
- During the day suck on ice cubes, frozen lemon or flavored water to encourage swallowing

When the first swallowing problems occur, it is not necessary to place the patient with Huntington's Disease on a mash diet. The patient can accept many foods in the normal way, sometimes with minor changes. Medication may be crushed and mixed with pudding or purée. Take into consideration that some medicines cannot be crushed, as it may change the efficiency and bio-availability. It is important to consult the prescribing doctor.

6.7. Recommendations for caregivers

- ✓ Ask your physician about a speech-language pathologist to obtain proper therapy and diagnosis of your swallowing disorder.
- ✓ It is useful to obtain a comprehensive list of appropriate food items for patients with swallowing disorders.
- ✓ The patient should be seated as straight as possible, preferably 90 degrees in a chair or wheelchair, not in bed, when eating.
- ✓ A quiet room without distractions is suitable for dining, and conversations with the patient should be avoided when the patient is eating or drinking.
- ✓ Feeding should be slow and served in small portions that are easy to chew and swallow. Patience is important, as you need to wait until food is swallowed before you put another piece in the patient´s mouth.
- ✓ Feeding must stop immediately, if the patient is coughing, choking or experiencing breath shortness. You should ask him to swallow several times without food in the mouth in such case. Always listen carefully to the voice quality; if you hear wheezing or hoarseness, stop feeding and wait.

- ✓ If the patient is bedridden, it is better to use a teaspoon instead of a tablespoon for eating to slow down the rhythm of the food considerably. During the meal, it is better to put only small food amounts in the patient's vision.
- ✓ The meal should not be interrupted, particularly in case of patients with an advanced stage of illness, as they cannot concentrate on the meal over a longer time. It may lead to irritation, frustration and aggression.
- ✓ Do not let the patient ingest solid food with fluids; it may cause inhalation and subsequent aspiration with choking.

Summary

- Dysphagia significantly increases the risk of choking, pneumonia, malnutrition and dehydration.
- Clinical signs of dysphagia are summarized in table 6.
- Aspiration symptoms are summarized in table 7.
- Silent aspiration occurs in patients with coughing reflex weakness or absence, when fluids and food enter the lungs.
- For swallowing disorder management follow chapters 6.2.2 and 6.7.

7. Dietary advice for Huntington's Disease

by Lucie Růžičková

The diet should be varied, well-balanced and nutritionally valuable. An insufficient intake of food may be noticed by unwanted weight loss or loose clothing. A patient's appetite can be increased by a glass of beer or a small appetizer before a meal. Strong meat or fish broth can as well increase the appetite. Important factors are also pleasant surroundings, a well-ventilated room and always enough time for the meal.

7.1. Recommendations for people without swallowing disorders

7.1.1. Basics about balanced diet

- ✓ Daily food consumption should consist of several smaller portions, preferably 5 – 6, considering breakfast, snack, lunch, snack, dinner, second dinner
- ✓ The interval between meals should not be longer than 3 hours
- ✓ Foods should contain an amount of energy to prevent unwanted weight loss, but not to cause an additional weight increase
- ✓ The diet should contain enough of full-value proteins, which are mainly found in lean meat, milk and milk products
- ✓ Various vegetables and fruits should be included daily into the diet
- ✓ Adequate fluid intake is important

7.1.2. Selection of suitable food

7.1.2.1. Meats and fish

It is important to consume more lean proteins, such as poultry, beef, pork, etc. At least twice a week one should include saltwater or freshwater fish. Preserved fish, e.g. sardines, smoked herring and salmon are especially encouraged. Sausages are also a good source of protein; one should eat poultry or pork/ham products with a higher proportion of meat, as well as lean sausages. Smoked meats and sausages contain a higher amount of salt, however, and should be eaten sparingly.

7.1.2.2. Eggs, milk and milk products

Eggs, milk and milk products are rich sources of proteins. The active lactic acid bacteria (probiotics) that are found in yoghurt, sour cream, kefir, buttermilk and quark have a beneficial effect on the body. Cheese is appropriate to include in the diet, not only

because of its high content of protein, but as well because of calcium, which is important for healthy bones and teeth. Hard and natural cream cheeses are preferred.

7.1.2.3. Fats

The most suitable are high-quality vegetable oils, especially canola and olive oil to use for cooking and preparation of cold dishes and salads. Butter and margarine may be used for bread and pastries or for the preparation of spreads.

7.1.2.4. Vegetables and fruits

Vegetables can be eaten cold, boiled, steamed or grilled and should be part of every main meal. It is ideal to consume a total of 500 grams of fruits and vegetables per day, with a ratio of 3:2. Vegetables contain a minimum quantity of energy and a high amount of fibre, vitamins and minerals. Fruits should be eaten as an early snack or afternoon snack.

7.1.2.5. Legumes

Legumes contain large amounts of fibre, which can cause bloating. Legumes are appropriate to use for soups, stews and porridge.

7.1.2.6. Pastries, breads and cereals

The best way to choose bread and pastry depends on the patient´s tolerance, whole grains being more valuable and nutritious. If whole grain breads cause irritation when swallowing, it is better to choose rather plain bread.

7.1.2.7. Spices

If one does not follow any dietary restrictions, one can use all kinds of spices that are tolerated well and do not cause irritation when swallowing. Fresh herbs- parsley, chives, rosemary, sage, stalk celery, dill etc. are as well, not a problem.

7.1.2.8. Drinks

It is advisable to drink regularly throughout the day. One should not drink large amounts of fluids before meals. If the patient is underweight, a good way to increase caloric intake is by sweetening drinks with honey or brown sugar. We need more fluids on hot summer days. Reduced fluid intake may cause decreased appetite.

7.1.2.9. Meal preparation

Meals should be prepared by boiling, stewing, and baking. Frying requires a preparation with a high amount of fat. One should eat fried food only once a week.

7.1.2.10. Menu suggestions

Day 1:

- Breakfast: Whole grain bun with butter, Gouda cheese and grated carrots
- Snack: Fruit salad or piece of fruit
- Lunch: Broccoli soup, pork with cooked vegetables and potato gnocchi
- Snack: Cottage cheese or quark with apricots
- Dinner: Pasta with ham and cheese, cucumber salad
- II. Dinner: Full-fat vanilla yoghurt

Day 2:

- Breakfast: Bread with cream cheese, chives and lettuce
- Snack: Baked apple with honey
- Lunch: Pea soup, beef with steamed rice and carrot salad
- Snack: Natural yoghurt with a banana
- Dinner: Whole wheat bread with fish spread and spring onions
- II. Dinner: Tomato juice

7.2. Recommendations for people with swallowing disorders

If one is experiencing problems while swallowing or chewing the food, the consistency of the meal can be adjusted into softer, ground or puréed form. In this case, it is necessary to ensure that the meal contains an adequate amount of all nutrients. A diet that is adjusted by mixing, usually must be diluted with a liquid, for example bouillon, broth, juice, milk or cream, depending on the type of meal. Nutrients are diluted as a result of this process. However, the meal amount increases in volume, making it even more difficult to consume, so it is difficult to maintain an adequate calorie level. Therefore, it is important to dilute the meal only with the smallest necessary amount of liquid and for preparation, to choose only nutritionally valuable foods, e.g. whipping cream, full-fat dairy products, butter, etc.

7.2.1. Basics about a puréed diet

- ✓ The daily menu consists of several smaller portions, preferably 5 – 6, considering breakfast, snack, lunch, afternoon snack, dinner, second dinner
- ✓ The interval between meals should not be longer than 3 hours
- ✓ The dishes included in the daily menu should have typical flavours and added seasoning should be well tolerated
- ✓ According to the condition of the person, adjust the food consistency to a uniform texture that is "spoon thick" like purée (or like apple sauce, hummus, pie filling, etc.) or to the smooth consistency of syrup " straw like".

- ✓ Meals with a puréed consistency should be served nicely on a plate, because visual appearance and sensory tasting is the key to acceptance of puréed food
- ✓ Foods may be puréed using a mixer / blender or food processor
- ✓ The daily diet should be nutritionally full with sufficient energy, protein, fat and carbohydrates amount
- ✓ If it is not possible to cover the energy requirements, the diet must be supplemented in another way, e.g. by sipping
- ✓ Even small amounts of food should have enough calories
- ✓ While eating, one should not drink a bigger quantity of liquids to avoid becoming full and getting an earlier sense of satiety
- ✓ The food's consistency must be adjusted to suit the patient´s individual needs and possibilities
- ✓ For more information, one should consult a nutritional therapist

7.2.2. Selection of suitable food

7.2.2.1. Meats and fish

Meats should be soft cooked, then puréed with smaller quantities of meat juices or sauce. Suitable are also meat casseroles which are prepared from minced meat. To increase the nutritional value, adding a small quantity of butter to the meat mixture is recommended. The meat will then be softer and thus easier to digest. Fish should be regularly included into the diet; boneless fish is fine in consistency and, therefore, very appropriate. Smoked meat and sausages should be prepared by fine mincing or slicing and may be added to cream cheese, cottage cheese, scrambled eggs, and mashed potatoes. Ham mousse is as well very tasty.

7.2.2.2. Milk and milk products

Milk is suitable for dilution of a purée, preparation of puddings, creams and mousses, hot chocolate/chocolate milk or white coffee. Whole milk or cream can be used to increase calorie intake, when necessary. As morning snack or afternoon snack, pudding, white yogurt, fruit yogurt without grains and cereals, cottage cheese, curd cream, kefir and other fermented milk products are appropriate. Cheese is a major source of protein. Grated hard cheese may be added e.g. into spreads or mashed potatoes. Cream cheese and cottage cheese may be used in different ways, e.g. with steamed vegetables, in soufflés or mashed potatoes.

7.2.2.3. Eggs

Eggs may be used for the preparation of a delicate custard, scrambled eggs or egg spread.

7.2.2.4. Fats

The most suitable are high-quality vegetable oils, especially canola and olive oil to use for cooking.

Butter is mostly added to soften the finished dish, or into spreads or mashed potatoes.

7.2.2.5. Vegetables and potatoes

Vegetables should be peeled, without tough parts and cooked tender and then mashed or puréed. Through the combination of different vegetables, one will achieve a variety of different flavours such as stewed spinach, steamed mashed carrots, boiled peas with melted butter, steamed broccoli with cream cheese, pumpkin purée, cauliflower custard, mashed peeled tomatoes, or tomato paste. Tomatoes can be easily peeled after steaming in hot water, and then halved to remove the seeds and the rest of pulp can be mixed or puréed. Potatoes must always be used in soft and mashed form for a fine consistency, and with the addition of celery or carrot purée, sour cream, cream cheese or mixed ham, different tastes can be achieved.

7.2.2.6. Fruits

Fruits should be ripe, peeled, free from tough parts and seeds, and then sliced, mixed, mashed or puréed. Bananas maybe easily mashed with a fork and mixed with yoghurt. Blended fruit compote or ready mixed fruits snacks such as apple, pear, apricot, or peach are very easy to digest. Fresh fruits such as peeled peaches, nectarines or apricots may be crushed or puréed. Fresh fruit juices can be prepared from various fruits; they should be diluted with water, because they may have a laxative effect in their pure form.

7.2.2.7. Legumes

Legumes contain large amounts of fibre, so they are necessary, if they are peeled and mixed very properly. Beans are appropriate to use in soups and purées.

7.2.2.8. Pastries, breads and cereals

The best way is to choose bread and pastry depending on the patient´s possibilities. In some cases, it is sufficient to get rid of the hard bread crusts. Another possibility is the inclusion of pre-soaked plain bread or milled rusk into spreads. Minced plain biscuits can be mixed into puddings, creams, yoghurts or mixed fruits or compotes. Rice should be mixed smoothly after cooking. Very small pasta noodles are best, e.g. vermicelli

should be chosen as it is easier to mix. To diversify the menu, it is recommended to take advantage of corn porridge (polenta), semolina, or millet (kasha).

7.2.2.9. Spices

If one does not follow any dietary restrictions, one can use all kinds of spices that are tolerated well and do not cause irritation when swallowing. Fresh herbs - parsley, chives, rosemary, sage, stalk celery, dill etc. have to be mixed or chopped into very fine pieces.

7.2.2.10. Drinks

The drinking regime is very important. A reduced fluid intake may cause a decreased appetite. In case problems occur while swallowing, thickeners should be used to achieve a proper consistency for the individual needs of patients. Sparkling drinks are not suitable for thickening. It is better to choose drinks with stronger aromas, such as pure fruit juices diluted with water.

7.2.2.11. Meal preparation

For the preparation of a puréed diet, it is the best to use already processed food by cooking, stewing or baked without any hard parts. Meat should be baked in foil for a better taste and to stay juicy and suitable for puréeing.

The food or liquid can be thickened with special thickeners of natural gums to achieve a better consistence for swallowing; thickeners may be added to hot and cold drinks or food. The quantity of thickeners should be controlled by the required consistency based on individual abilities of the patient. Thickeners are without flavour, and therefore do not alter the taste of food or beverages.

7.2.2.12. Example menu

Day 1:

- Breakfast: Porridge (from fine flakes) with chocolate powder, maple syrup and mashed banana
- Snack: Fruit snack with cream cheese
- Lunch: Pea soup, pork mixed with meat sauce and oregano, potatoes mixed with celery, parsley, carrots and cream with tomato sauce (from peeled, seedless and chopped tomatoes)
- Snack: White yogurt with melted chocolate, garnished with cinnamon and honey
- Dinner: Vermicelli pasta baked with spinach and cheese, puréed peach compote
- II. Dinner: Ready-made cream cheese dessert

Day 2:

- Breakfast: Cheese spread with a bread roll soaked in milk
- Snack: Vanilla pudding
- Lunch: Cauliflower cream soup, baked fish fillet in foil with lemon and butter with mixed rice pudding and beetroot
- Snack: Mixed apple compote
- Dinner: Corn porridge (polenta) with blue cheese
- II. Dinner: Tomato juice Summary

- It is important to eat regularly, 5-6 times a day.
- Ensure a sufficient supply of energy and protein
- To improve the appetite, treat yourself with a small glass of beer or a small aperitif
- The puréed diet should be tasty and prepared from nutritionally valuable foods, e.g.
- whipping cream, full-fat dairy products, butter, etc.
- It is important to dilute the puréed or mixed meal only with the smallest necessary amount of liquid
- There must be enough energy even in small amounts of food
- Thickeners should be used to achieve a proper consistency for the individual needs of patients

8. Clinical nutrition and its administration

by Jiří Klempíř

8.1. Basic definitions

Normal diet is defined as food that the individual eats in everyday life and is as well served in sanitary facilities.

Diet as a word describes the use of specific nutrition methods to improve one's health or well-being. The different types of diets may vary in energy content or in amounts of nutrients.

Nutritional support is therapy for patients who are suffering from malnutrition and cannot get enough nourishment by eating or drinking. It is a concerted effort to improve health through a fortified diet, or enteral or parenteral nutrition.

Fortified foods differ from a normal diet due to their higher energy content and higher amount of micro- and macronutrients (see chapter 3.2.4.). Food fortification or enrichment is the process of adding micronutrients (essential trace elements and vitamins) to food.

Clinical nutrition is a special diet that helps to improve the patient´s health and to enhance the body's processes for a better and more effective treatment.

Enteral nutrition applies to any method of feeding that delivers nutrition to the alimentary tract (gastrointestinal tract), it includes an oral diet (e.g. sipping) as well as tube feeding with a nasogastric feeding tube (figure 5) or with a gastrostomy feeding tube (figure 6, figure 7).

Parenteral nutrition refers to the supply of nutrients through the blood.

8.2. Sipping

If the patient has no significant problems with chewing and swallowing, he may use artificial nutrition in the form of high-energy drinks and puddings (e.g. **sipping**, means of drinking a liquid diet. It is based on a complete formula of nutrients, which differ from each other based on calorie level, the number of macronutrients and micronutrients, and fibre. **Most of these products do not contain lactose or gluten**. The energy content per unit of volume is at standard preparations 1 kcal /ml; there are as well products with a higher energy content (1.25 – 2 kcal /ml.

Sipping is mostly enjoyed in addition to the normal diet, between meals or as a supplement to the diet. Sipping should not lead to a reduced intake of normal food.

The products usually have a sweet taste, but there are also salty ones or ones without flavour, as well as in the form of juice (without fat.

Diabetic patients with poorly controlled diabetes should use products with lower energy and carbohydrate levels that do not impact fluctuations in blood glucose. Standard products or products with higher energy may be used to combat malnutrition in the case of patients with manageable diabetes.

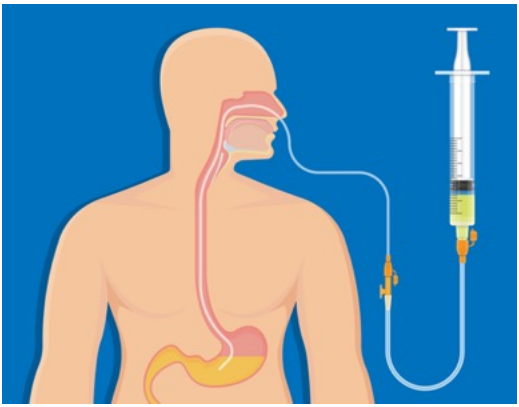
For patients with fluid dysphagia (swallowing problems when drinking beverages, liquid products for sipping, sauces, and soups it is recommended to use commercially produced thickeners based on modified starch or natural gums, which do not affect the taste of drinks.

If the patient is unable to orally consume a sufficient amount of nutrients or there is an obstacle in the alimentary tract on its way into the stomach, a feeding tube with an alternative access to the stomach has to be used: a **nasogastric feeding tube** (figure. 5) or **gastrostomy feeding tube** (figure 6, figure 7).

8.3. Nasogastric tube

The **nasogastric tube** (NGT) is a plastic feeding tube that goes through the nose into the stomach for administration of nutrition (figure 5) This method works for short-term use: no longer than 2-3 weeks. The long-term use of a nasogastric tube may damage the mucosa of the upper respiratory tract and cause pressure sores.

Figure 5: Nasogastric tube (NGT)



8.4. Percutaneous endoscopic gastrostomy (PEG tube)

The **percutaneous endoscopic gastrostomy** (PEG) tube, also known as gastrostomy tube or gastricfeeding tube (G-Tube) is a solution for nutrition administration over a longer period. The tube is placed into the stomach under endoscopic control, through a small puncture of the front wall of the stomach and affixed onto the abdominal surface while the patient is under mild sedation (figure 6).

Figure 6: Insertion of a percutaneous gastrostomy tube (PEG)

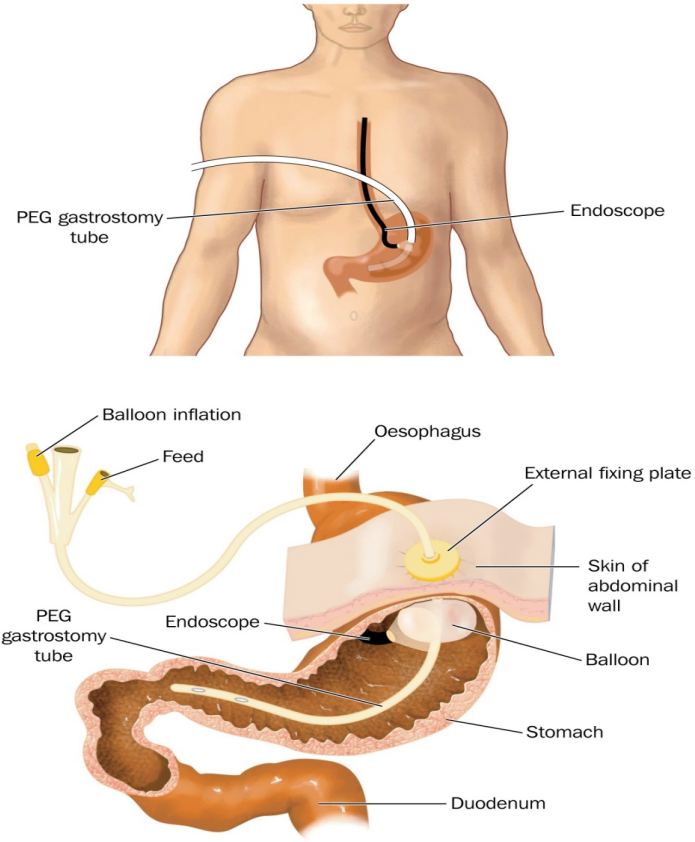
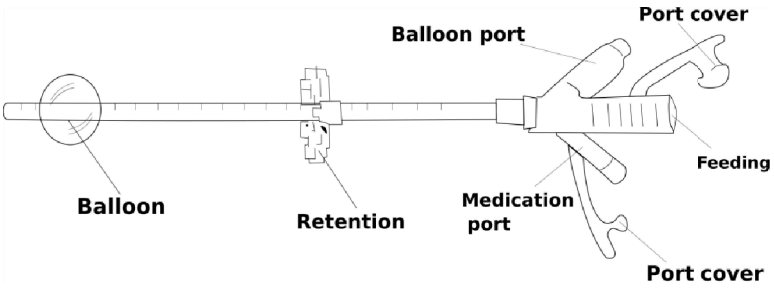


Fig. 7: Percutaneous gastrostomy tube (PEG)



The implementation of a PEG does not necessarily mean the patient is unable to receive food and fluids by mouth. Ideally, PEG is used in combination with the intake of nutrients orally for better food enjoyment and in order to achieve an adequate and balanced nutrition and fluids intake.

Patients, their relatives, and caregivers often have an unexplained exaggerated fear of PEGs and consider this method to be a complication and indicate the diseases' progression. Actually, a correctly placed PEG may prevent complications, facilitate feeding, and reduce the risks associated with food intake in patients with dysphagia (e.g. pneumonia, suffocation).

On the other hand, the implementation of a PEG in a patient with severe malnutrition or in patients in critical condition is associated with complications. It is contraindicated to administer a PEG in terminal stages of illness when death is expected within days or weeks.

If there are no reasons for an enteral feeding via PEG and the condition of the patient has improved, the PEG may be easily removed by endoscopic surgery. The opening in the abdominal cavity is usually sealed within 24 hours and leaves a small scar.

8.5. Parenteral nutrition

Parenteral nutrition is administered into the bloodstream in the form of an infusion, when the digestive tract cannot be used temporarily for nutrition intake. In patients with Huntington's disease, this method is used only for a short period in exceptional cases when it is not possible to administer diet safely into the digestive tract (e.g. acute abdomen).

8.6. Comparison of parenteral and enteral nutrition

Based on clinical experience, the preferred form of nutrition is the enteral one, because it keeps the function of the gastrointestinal tract, it improves immunity, it is easier to apply in the home environment, it is comparatively inexpensive and finally, it is associated with fewer complications for the patient. Enteral nutrition and fluids via feeding tubes (NGT or PEG) may be administered by bolus or continuously by an enteral syringe pump (8 to 22 hours per day).

- Enteral nutrition is clinical nutrition with an exact composition of nutrients
- (carbohydrates, fats, proteins, minerals, trace elements, vitamins) administered via the digestive tract
- Sipping (nutrition for drinking) is intended for patients with a good functioning gastrointestinal tract, who are unable to maintain an adequate body weight through a normal diet
- If the patient is able to safely consume a sufficient amount of food and energy orally, he / she does not need sipping
- Percutaneous endoscopic gastrostomy (PEG) is an implantation of a thin tube
- (feeding tube) through the front abdominal wall into the stomach for feeding
- PEG is intended for patients who cannot safely and in sufficient quantity take nourishment orally
- The introduction of a PEG tube does not automatically mean the termination of eating by mouth
- The implantation of a PEG tube in proper times prevents complications, and improves the nutritional status and quality of life
- If a PEG tube is no longer necessary to maintain adequate nutrition, it can be kept or removed, and the wound heals leaving a small scar
- The implantation of a PEG tube for patients in critical condition or at terminally advanced stages of disease is unethical and not recommended

9. Management of PEG tube

by Petra Havrlíková, Martina Dvořáková

Percutaneous endoscopic gastrostomy (PEG) is in fact nothing more than a thin tube for feeding, of which one end is brought into the stomach and the other end to the skin of the abdomen (chapter 8.4.). This allows administration of nutrition directly into the stomach and prevents leakage of nutrition beyond the stomach.

9.1. Taking care of PEG in the first seven days after insertion

The hospital stuff will take care of the implanted PEG feeding tube in the first two days, at the same time, the patient should learn how to take care of the tube

After discharge from hospital, in the first seven days it is necessary to follow a daily sterile care of the PEG tube. The patient should check the place of insertion and its surroundings for bleeding, redness, allergic skin reactions, hardening around the tube, pus, and nutrition leakage.

The following steps are recommended:

(1) Proper hand hygiene disinfection is very important before daily bandage change is done, the disinfection agent can be purchased at a pharmacy or drugstore.

(2) First you carefully release the blue/yellow tubing clamp by gently pulling, followed by releasing the silicon clips of the external silicon bumper (figure 8, figure 9).

(3) Then you slide the external silicon bumper by moving it 5 cm outwards from the abdominal wall. You should properly clean and pat dry the place of the tube insertion and the outer bumper from both sides. It is very important to keep the insertion place and surrounding skin clean and dry. Do not use the following agents for disinfection: Povidone-iodine, also known as iodopovidone (Betadine, Braunol, Braunovidon, Jodisol) and octenidin (Octenisept). Those agents may adversely affect physical and mechanical properties of the PEG tube.

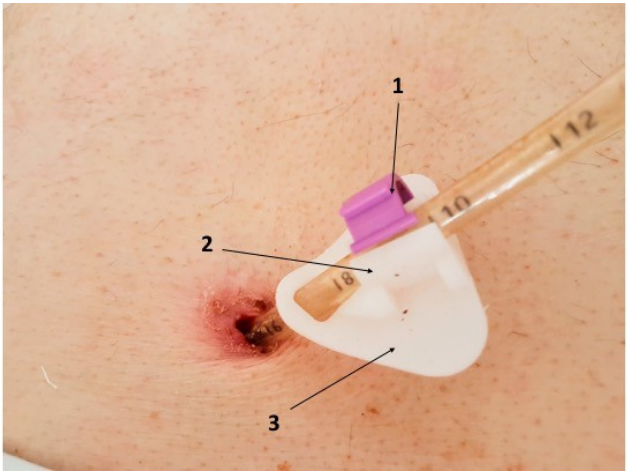
(4) In the first seven to ten days, the sterile square gauzes should be placed underneath of the outer silicon bumper or crossbar.

(5) After the skin and place of insertion is dried properly, the outer bumper can be moved back to its place and fixed by the clamps. At the end the tubing clamp should be slide back and as well fixed. Between the outer bumper and

the abdominal skin should be a space of about 5 mm to avoid pressure sores (figure 10). You can check on the proper placement of the tube by gently pulling on it, the external bumper should not leave marks on your skin. A violent tube resection could lead to removal of the internal disk from the stomach into the abdominal cavity and might damage the gastric mucus

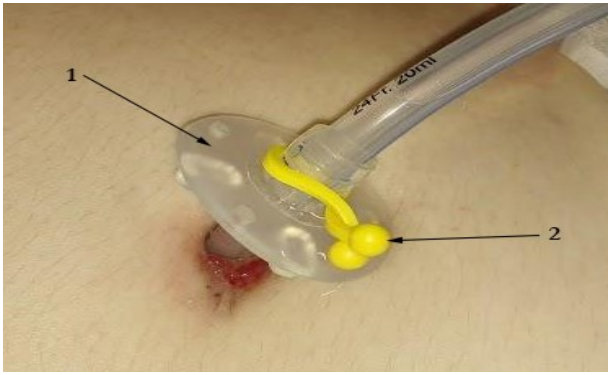
(6) The end of PEG Tube can be rest gently on the abdominal wall or be fixed by dressing, if needed (figure 11).

Figure 8: PEG tube



1 - tubing clamp, 2 - clamps of outer bumper, 3 - outer (external) silicone bumper

Figure 9: PEG Tube – different type



1 - external silicon bumper/ crossbar, 2 - external circle clamp

Figure 10: Position between abdominal wall and outer bumper

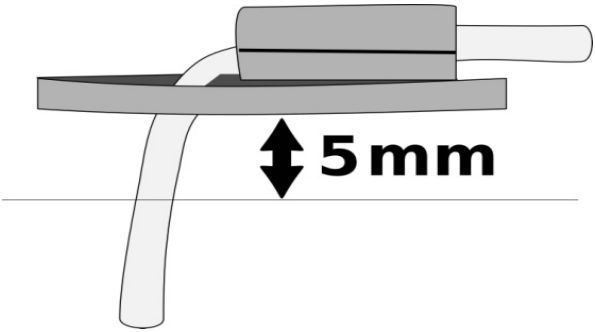
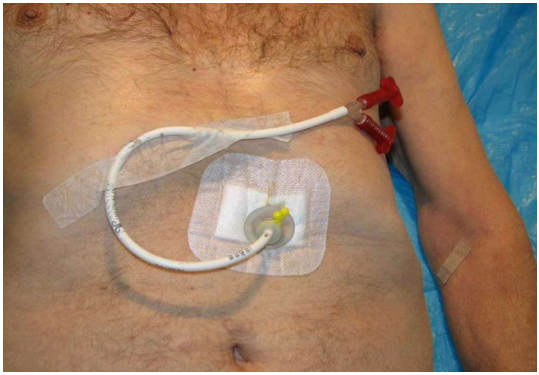


Figure 11: Dressing of PEG tube



Summary

- It is recommended to take care of the PEG tube during the first seven days after placement in accordance with point 1 to 6.
- In case of questions, you should always contact your physician or hospital, where the PEG tube was administered.

9.2. Taking care of the PEG in long term

PEG treatment in daily care:

(1) Before handling the PEG tube pay attention to hand hygiene.

(2) Release and slide the external silicon bumper 5 cm outwards from the abdominal wall.

(3) Push the tube 2 - 3 cm into the stomach (figure 12) and rotate the tube through 360° (figure 13). It is important to rotate the tube gently each day to prevent scar tissue from forming.

(4) After returning the tube to its original position, carefully rinse the PEG tube and external bumper from both sides, with wet gauze pads, for example, then dry properly.

(5) At the end the outer bumper should be moved back and fixed as usually, it is important to leave about 5 mm between the outer bumper and the abdominal skin to prevent pressure sores

(6) Each day pay attention to the end of the tube and clean it, e.g. with a toothbrush.

Figure 12: Mobilisation of PEG Tube

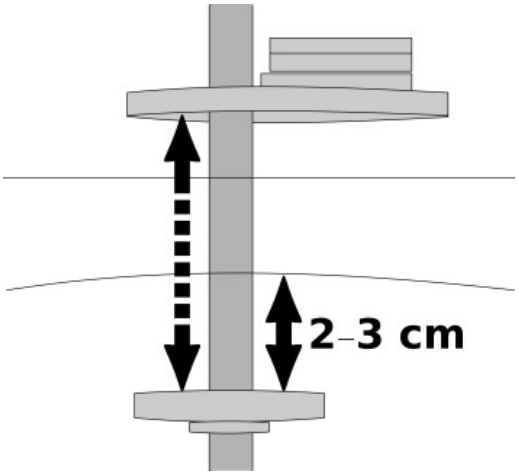
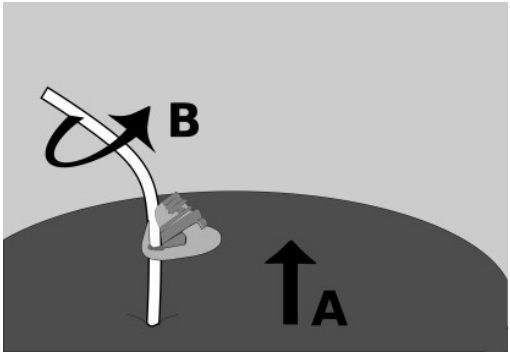


Figure 13: Rotation of PEG Tube



Summary

- In daily care of the PEG Tube follow the steps 1 to 4.
- In case of questions, you should always contact your physician or hospital, where the PEG tube was administered.

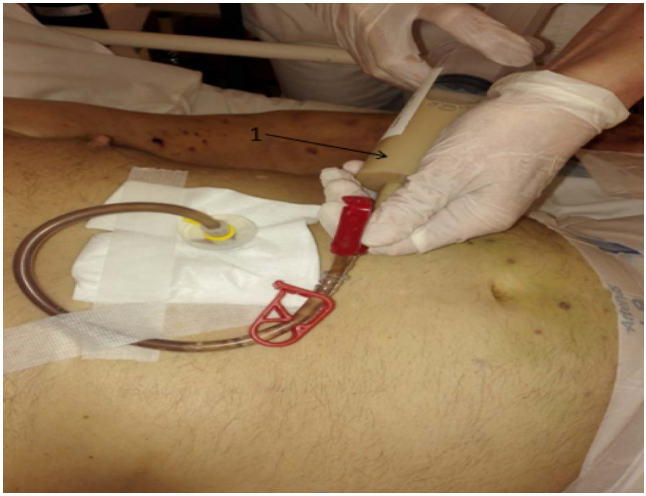
9.3. Administration of nutrition via gastric tube

Only nutrition formulas designed for PEG tubes should be administered according to the recommendation of your physician or dietician. Regular food, alcohol, fruits, or other foods in mixed form are not recommended. Also, liquids with acid content could lead to a shrinkage of nutrition and obstruction / blockage of the tube.

- ✓ In the first 24 hours after tube placement, water should be administered, when well tolerated, and followed by continues slow administration of nutritional feed. The first nutrition should be given as a bolus dose of 50 ml, when well tolerated, followed by a continuous dose increase (approximately about 50 ml per dose) to achieve about 250 - 300 ml per dose.
- ✓ Nutrition should be administered by special syringes, e.g. purple syringe in the United Kingdom (figure 14). One syringe is needed for nutrition and a separate one for feeding tube rinsing. It is important to rinse the tube with 50 ml purified water or boiled cooled tap water after each nutrition administration.
- ✓ Intervals recommended between single dosages are about 2 - 3 hours. The dietitian or physician will write a diet plan with the exact dosage of nutrition formulas and water. In case the patient is not drinking any fluids orally, you should administer around 150 - 200 ml of water between single nutrition portions.
- ✓ The open nutrition should be stored in the refrigerator, approximately one hour prior to administration it should be taken out.
- ✓ The nutrition should not be administered during the night.
- ✓ After completion of the feeding, the feeding tube has to be closed properly to prevent leakage of nutrition or gastric juices. If the feeding tube is not used on a regular basis, it is important to rinse the tube daily with 50 ml water.
- ✓ The patient should be in sitting, or at least in half-sitting position during feeding and remains in this position at least 30 minutes after application for better digestion and tolerability.
- ✓ If prior to the administration the gastric contents or previously administered formula is coming out of the feeding tube after opening, or the patient presently feels a sense of fullness and satiety, it is necessary to postpone the administration of the next nutrition formula.

- ✓ Proper daily oral hygiene is necessary even if the patient is not receiving nutrition and fluids by mouth.
- ✓ Another option for tube feeding is the use of an electric pump to control the flow of the formula into your PEG tube. Your dietician will teach you how to set up and use the pump.

Figure 14: Formula administration via syringe for PEG tube



- During the nutrition administration and for at least 30 minutes afterwards a proper position of the patient in sitting up or half-sitting (if tolerated) is very important.
- If the patient does not receive any fluid orally, the fluid administration should be given regularly.
- PEG equipment has to be kept clean and dry.
- Every day oral hygiene is very important.
- Similarly, to the PEG tube, the nutrition could be as well administered via nasogastric tube (NG), tube introduced through the nostril to the stomach (chapter 8.4).

9.4. Administration of medication via the gastric tube

Due to individual patient's requirements, it is often necessary to administer medication via the PEG tube. Preferably the drugs should be used in the form of liquid preparations (drops, syrups) suitable for PEG, diluted with water.

In case of tablets, it is important to obtain information on whether the tablets are suitable for administration via feeding tubes, e.g. in crushed form. Some medication can change the bioavailability and its properties, when crushed or diluted, hence it is important to consult your physician.

If the drug is available only in tablet form, it has to be crushed and properly mixed with 15 -30ml of water. Every drug should be crushed separately. The feeding tube should be rinsed properly with water before and after drug administration.

The medication should never be added to nutrition formulas; this could cause a change of nutrition consistency (shrinkage) and feeding tube blockage.

Medication that should be taken on empty stomach should be administered before nutrition is served, other drugs should be given about 30 minutes after feeding.

Summary

- Before any drug administration via PEG tube consult your physician.
- Always rinse the feeding tube properly with water before and after drug administration.

9.5. PEG Complications in everyday life and its solutions

9.5.1. Obstruction / blockage of the gastric tube

If the patient´s feeding tube is blocked, it should be flushed with warm water (not hot) using the small size syringe (10 or 20ml). It is also suitable to use slightly heated mild carbonated water. Flushing should take place in short intervals and in small doses, approximately 10 ml, the procedure should be repeated.

If not successful, the following procedure can be tried: One tablet of pancreatic enzyme (Viokase, Creon, Panzytrate, purchased at the pharmacy) should be crushed and diluted with water and immediately injected into the tube and clamp, to remain about 5 - 10 minutes in the tube, and afterwards flushed away with water.

NEVER attempt to remove the blockage by mechanical force, or by inserting metal objects.

The physician should be contacted immediately (within 24 hours), if the blockage cannot be released.

9.5.2. Stoma leaking

It is necessary to check the place of tube insertion. If there is nutrition or gastric fluid leaking, you should contact the physician within 24 hours.

9.5.3. Diarrhoea, vomiting

Diarrhoea is when the patient has more than three loose or liquid stools per day. This may be caused by rapid administration of nutrition, serving excessively large single doses, short intervals between doses, unsuitable nutrition temperature, nutrition intolerance, or disorders of the intestinal microflora (e.g., after prolonged use of antibiotics).

It is important to observe and to try to identify the cause, if problems persist, the physician should be consulted.

9.5.4. Constipation

Constipation could be caused by administration of too little water or using an improper nutrition formula. For prevention, formulas containing fibres should be administered. The condition should be addressed during the next appointment with the dietician or physician.

9.5.5. Loss of gastric juices or nutrition formula

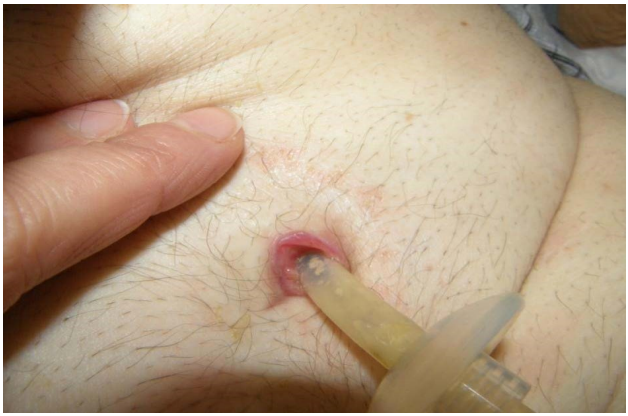
Prevention of leakage is very important by proper position during feeding, such as sitting or at least 45degree angle (half sitting) and to remain in this position for 30 -60 minutes following the end of the feeding. A proper nutrition dosage should be administered. The outflow of larger amounts leads to serious complications and the physician should be contacted immediately.

9.5.6. Granulation tissue

Granulation tissue is pink/red, cauliflower-like tissue, that grows around the tube in the place of insertion, it is very vulnerable and starts to bleed by minimal mechanical irritations (figure 15).

It can be caused by incorrect tube stabilisation in stoma, too large space (more than 5mm) between the abdominal wall and the outer bumper, or by excessive moisture around the place of insertion. The physician should be consulted at the next appointment.

Figure 15: Granulation tissue around PEG Tube

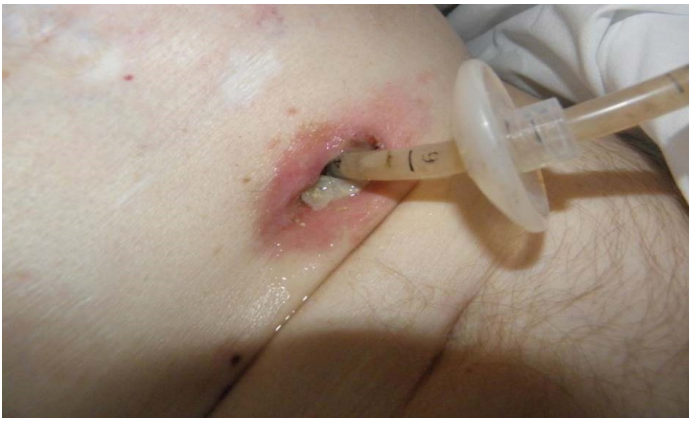


9.5.7. Skin redness and infection

Circular redness of less than 5 mm is considered normal and not necessarily a sign of infection, it is important to daily monitoring (figure 16).

The infected place should be treated twice daily and the sterile dressings with gauze squares, as in the first days after PEG tube placement, should be applied. In case of worsening, swelling, or fever you should consult your physician, who should apply a topical ointment and prescribe a local antibiotic agent.

Figure 16: Skin redness and infection



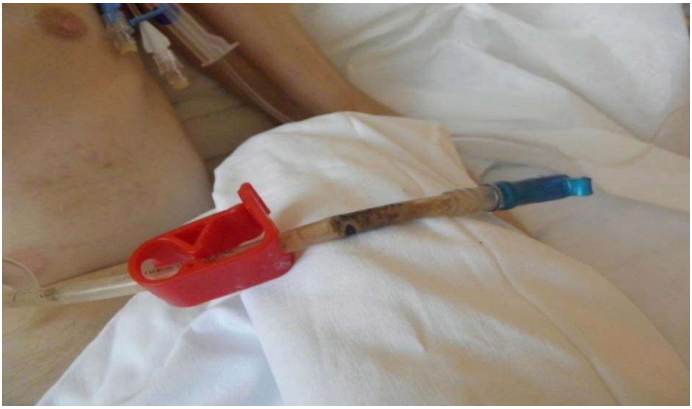
9.5.8. Tube dislocation / dislodgement

Accidental tube dislocation or dislodgment can happen. It is important to know that the fibrous channel in which the tube is located starts to rapidly retract / close (within hours). You should consult a physician immediately or go to hospital for replacement.

9.5.9. Rupture, break, damage of the PEG

Damage of the PEG tube occurs most often when the tube is in place and used for a longer period of time, or in case of rough handling (figure 17). Rupture or cracks may occur in locations where the tube clamp is located. For prevention, the clamp should be used on different places and over the whole length of the tube. In case of damage, it is possible to shorten the tube or a replacement has to be inserted by your physician.

Figure 17: Damage of the PEG

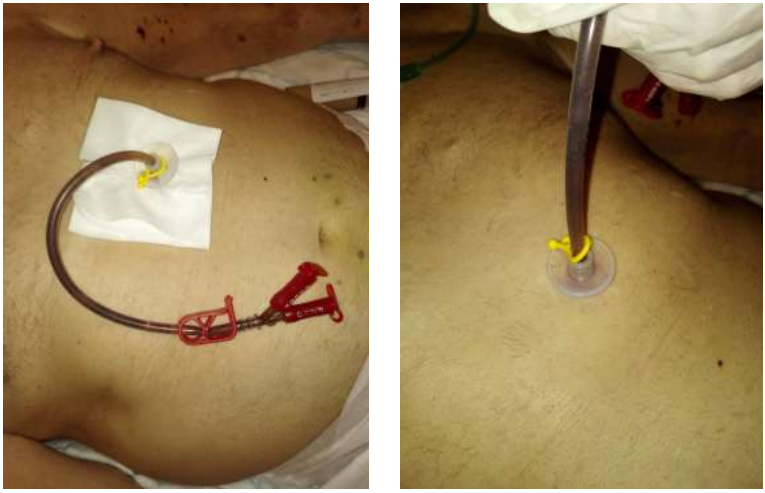


9.5.10. Dark colour in the tube

The dark colour of the PEG tube occurs most often when the tube is in place and use for a longer time, usually caused by flushing with strong black tea or aggressive fluids containing dye (figure 18).

Darker tube colour does not affect its function.

Figure 18: Dark colour in the tube



9.5.11. Buried Bumper Syndrome

It is a complication that occurs when the tube is in place and use for a longer time, when the inner tube plate overgrows by scar tissue within the stomach wall itself. For prevention, it is important to rotate the tube daily around 360°.

- If you are not sure how to deal with problems or complications, feel free to contact your attending physician.

10. Basic facts about medical nutrition supply

by Jiří Klempíř, Alžbeta Mühlbäck

Enteral nutrition formulas and equipment are supplied by a number of companies to pharmacies. Individual products may vary in the form of packaging, taste, consistency, concentration, composition, amount of fibre, energy content and price (chapter 8).

If a physician or nutrition specialist prescribes nutrition formulas or nutritional supplements for patients, the cost for the patient are significantly lower or even free of charge. In many countries, the enteral and parenteral nutrition is considered to be a drug (medicine, or food formulas for special medical purposes, chapter 11) and is at least partially covered by health insurance.

If the nutrition is freely purchased at the counter in pharmacies, in most cases, it is not entitled to reimbursement by the insurance company. The amount of prescribed nutrition, forms, and duration of administration is depending on the current health of the patient (chapter 5).

- If your physician, dietitian or nutrition specialist correctly indicates the treatment and use of clinical nutrition, the treatment will be covered by health insurance in most countries.

11. Food for special medical purposes and food supplements

by Jiří Klempíř, Alžbeta Mühlbäck

11.1. Food for special medical purposes

Medical products prior to introduction to the market in the EU must undergo an authorisation process from either a national authority or the European Medicines Agency. In this procedure, the quality, safety, effectiveness and therapeutic indications are proven and evaluated.

These products are different from ordinary foods, either by their special composition or a special manufacturing process and are intended for dietary management under medical supervision, e.g. sipping, nutrition for PEG tube. (chapter 8).

11.2. Food supplements

Food supplements are considered to be an additional source of nutrients and other substances to normal diet. The EU Directive 2002/46/EC specifies requirements for food supplements, their safety, but no curative effects. Therefore, dietary supplements cannot be considered a method of medical treatment.

Dietary supplements include: Vitamins, minerals, trace elements, extracts of medicinal herbs (e.g. guarana, ginseng, garlic), antioxidants, omega-3 fatty acids, amino acids, melatonin, various kinds of fibres, probiotics (chapter 3.2.6) and other substances (e.g. glucosamine, chitosan, choline, lecithin, carnitine, coenzyme Q10, creatine).

However, food supplements failed so far in patients with Huntington's disease to show any positive effect. It is unlikely that any of dietary supplements could reverse the course of Huntington's disease.

Summary

- Food for special medical purposes (e.g. sipping, nutrition for PEG tube) should be prescribed only by a nutritional specialist.
- Dietary supplements are freely sold and their effectiveness cannot be guaranteed, only their safety.
- These food supplements cannot slow or stop the progression of Huntington's disease.
- The patient should consult the physician prior to trying food supplements and in case of administration, evaluate its effects after 2 – 3 months of administration.

12. Basic facts about medications and nutrition in Huntington's Disease

by Jiří Klempíř, Alžbeta Mühlbäck

12.1. Basic information about medication use

It is not possible during the course of Huntington's disease to avoid using certain medications for symptomatic treatment. Each drug has its side effects, but under certain circumstances even "unwanted" effects became positive effects, such as improving appetite and weight gain. For each medication, there is usually a long list of possible side effects. Some are more common; others are quite rare. The possible presence of side effects depends on a number of factors such as the type and

stage of disease, the presence of other diseases, medication dosages and interactions with other drugs, and especially individual sensitivity to the substance.

In general, caution is required when treating children, elderly, when taking multiple drugs simultaneously (drug interactions) and in the case of severe heart disease, kidney, liver and pancreas problems. Alcohol consumption can as well cause occurrence or worsening of side effects.

It is generally possible to be prescribed a drug with the same active ingredient, but under different trade names, these are called generic drugs. They are equivalent in quality, dosage, route of administration, but may differ in price and packaging. This sometimes leads to unnecessary use of the same substance under different brand names. Therefore, the following list shows a short overview of active substances which are always listed on the packaging.

12.2. Antipsychotic

This group is mostly used for the reduction of choreatic movements. The most common substances are tiapride, risperidone, haloperidol, olanzapine, quetiapine, melperone, and more. Basically, these drugs reduce involuntary movements, but rarely can worsen the function of voluntary motor system and lead to disturbances of speech, swallowing, fine motor skills, walking and stability. Antipsychotics are as well used to treat irritability, verbal and physical aggression, hallucinations, delusions and delirium. Some of antipsychotics are used in management of nausea, vomiting and hiccups. A side effect may be increased appetite and weight gain.

12.3. Tetrabenazine

Tetrabenazine is used for the symptomatic treatment of hyperkinetic movement disorders. Attention is required by unusual changes of behaviour as it can lead to worsening or occurrence of depression and suicidal thoughts. Rare side effects on the digestive system include diarrhoea, constipation, and vomiting.

12.4. Amantadine

Amantadine is used for the reduction of dystonia, Parkinsonian syndrome, and stability disorders. Amantadine can cause dry mouth and urinary retention in relation to prostate enlargement. Adverse effects also include agitation, exacerbation of seizures or psychiatric symptoms.

12.5. Valproic acid

Valproate is known as antiepileptic drug and mood stabiliser. It is used in treatment of seizures and myoclonic disorder and as well against irritability and aggression. A positive side effect may be increased appetite and weight gain. Digestive problems such as nausea are rare and typically only last temporarily.

12.6. Antidepressants

Antidepressants are used for the therapy of depression, anxiety and sleep disorders and some substances even show a reduction in irritability and aggression. Untreated mood disorders and anxiety can lead to low appetite and weight loss. Some antidepressants can cause an increased appetite and body weight gain. Side effects such as paradoxical feelings of inner restlessness, insomnia and nausea, and diarrhoea may appear at the beginning of the administration of antidepressants treatment. All these symptoms normally disappear after adaptation time and the medication shows its positive effects.

12.7. Benzodiazepines

Benzodiazepines (clonazepam, diazepam, alprazolam, bromazepam, oxazepam) are used for the symptomatic treatment of anxiety, sleepiness and very intense involuntary movements. Their long-term use often leads to dependence syndrome. Side effects include the deterioration of speech and swallowing, lack of coordination, incontinence, drowsiness, sedation, respiratory depression, and impaired attention and memory functions. Adverse effects are potentiated in combination with substances that also show sedative effects including alcohol.

12.8. Antiemetics

Antiemetic drugs are effective against nausea and vomiting due to swallowing disorders. Some antiemetics (thiethylperazine, metoclopramide) cause similar side effects on the motor system as antipsychotics. Substances like itopride and domperidone show no adverse effects on the motor system. The latest antiemetic drugs (e.g. ondansetron, granisetron, tropisetron) are very effective in the therapy of nausea, though they can cause constipation, diarrhoea and headache.

12.9. Antacids and antiulcer drugs

Antacids neutralize stomach acidity and help to relieve indigestion. The most common antiulcer drugs are proton pump inhibitors (e.g. omeprazole, pantoprazole) that inhibit the gastric acid production that can irritate the stomach and oesophagus (heartburn, oesophageal or gastroesophageal reflux disease, ulcers). Their excessive or uncontrolled use may reduce the absorption of some food components.

12.10. Laxatives

Constipation is a digestive disorder that manifests with infrequent and slow bowel movements and the stool becoming hard in consistency. The cause can be wrong diet with lack of fibre, lack of fluid and movement or medication side effects. Treatment should be focused on the underlying cause. Constipation may lead to reduced appetite, bloated stomach and abdominal pain. In some cases, the use of laxatives cannot be avoided.

Bulk-forming agents such as dietary fibre increase the volume of the intestinal contents and soften the stool. Because they absorb plenty of water, sufficient fluid intake is required at administration. They are gentle in their effect and can be taken in long term use.

Hyperosmotic laxatives increase the volume of water in the colon, stimulating bowel movements (magnesium, lactulose, glycerol).

Stimulant agents act on the intestinal mucus, reduce the absorption of water and secondarily cause strong intestinal motility.

Frequent and uncontrolled use of laxatives may reduce their effectiveness. A laxative overdose may lead to diarrhoea and dehydration.

Summary

- Health status changes are not always side effects of treatment, but may also be signs of a health problem that is not related to Huntington's disease, or may be part of the underlying disease's progression.
- The patient should always consult their doctor prior to any medication´s change.

13. Physiotherapy in Huntington Disease

by Romana Konvalinková

Physiotherapy is an important part of care and management of patients with Huntington's disease. Physiotherapy should be automatically integrated into the daily routine, from the very beginning, respectively, from the time of Huntington´s disease's diagnosis. During the course of the disease, physical symptoms (chorea, rigidity, impaired voluntary movement) are present and as well behavioural disorders such as personality changes, decreased cognitive performance, especially memory and concentration difficulties, and apathy (chapter 2). The interaction of different above mentioned symptoms may lead to the fact that motivation to exercise is not always given and therefore, patients need support and additional motivation from their family, friends, and caregivers.

13.1. Goals of physiotherapy

The aim of physiotherapy is to reduce gait disturbance with impaired stability leading to falls, optimize the impaired fine motor skills, increase the reduced muscle strength, and relieve fatigue. The physiotherapist in collaboration with the speech language pathologist also concentrates on swallowing disorders (chapter 6) and breathing exercises. Physiotherapy focuses on the maintenance of active movements and independence. In advanced stages, the strengthening of respiratory muscles, including support of coughing reflex and expectoration, and maintenance of passive joint movements as a prevention of spasticity and bedsores, is the primary concern. Exercise generally prevents excessive blood clotting, bedsores, and improves cardiovascular circulation, respiratory functions, immunity and psychological condition.

13.2. Physiotherapy and nutrition

Physiotherapy is focused on the maintenance of a functional muscle mass and independent physical condition. **Malnutrition contributes to loss of muscle function** and is caused by various factors (chapters 5.2, 5.4 and 6). The organism obtains energy and nutrients during the malnutrition state by degradation of its own skeletal muscles (chapter 5). Involuntary movements themselves, unless they are very intense, are not causing high-energy consumption. Conversely, **apathy is worsening** with the progression of Huntington´s Disease, that **leads to a reduction of overall motor activity and thereby reduction of muscle mass**. The **patient perceives** this **state as tiredness, exhaustion and weakness of skeletal and respiratory muscles**. Patients in advanced stages of malnutrition (even without serious motor system dysfunctions) have

problems with self-care, self-motivation and tolerance of exercise. Slowly and continuously a vicious cycle develops, which is difficult to break through.

A sufficient energy supply is always necessary to combine with physical activity, since exercise stimulates protein synthesis and a proper functioning of the muscle mass. Generally, this simple rule applies to both groups, healthy people and people with Huntington´s disease. Therefore, **muscle areas in need of strengthening need to be targeted regularly by training**.

13.3. Physiotherapy in different stages of Huntington´s Disease

In the early and middle stages of the disease, **aerobic exercise** is recommended at least three times a week for 30 - 45 minutes. It is any exercise, that the individual enjoys with involvement of large muscle groups, performed at moderate intensity (increased respiratory rate, but one is still capable of conversion) and increased heart rate (55 – 80 % of the maximal heart rate). Examples are running, Nordic walking, walking as well without poles (if disturbed by choreatic movements), swimming, biking or treadmill.

The **strength training** of large muscle groups is recommended at least two times a week to maintain the muscle strength or even, if necessary, to increase the functional muscle mass. The common exercises for lower limbs include: standing calf raise and pulse, squatting, side and forward lunges, repeating sitting and standing up, and step ups by using a platform or steps. It is possible to use different weights for exercising the upper limbs, or to use the weight of one's own body (push ups).

It is important to focus on **balance and core stability training**, such as lunges in all directions, squatting, balancing on one leg, standing on balance board or soft pad etc. During the exercise safety has to be ensured and injuries and falls should be avoided (practice against the wall, with railings etc.)

An integral part should be training while carrying out multiple tasks (e.g. training and also to count or solve logical tasks, to name the words of one letter etc.).

Different exercises may be combined in the form of **circuit training**, which is efficient, but has to be performed on a regular basis (figure 20).

Figure 20: Aerobic training

13.4. Physiotherapy programs in advanced stages

During **advanced stages** of Huntington´s disease, it is important to take care of daily active exercises depending on the patient´s physical condition, whether standing (figure 22), sitting (figure 23) or in bed (figure 24). It is important that the family, caregivers or nursing stuff are aware of daily training and able to support it. The best way is to start early by developing strategies for mobility maintenance and daily living activities. The possibility of compensatory aids should not be forgotten and offered to the patient, if needed.

Part of the daily training should be a regular stretching of the muscles at risk of stiffness.

Stretching should be performed within the maximum possible range of motion and stay in the stretched position for a few minutes. Stretching should be always done in pleasant positions that the muscle pulls but does not hurt! Stretching prevents muscle

and soft tissue changes, improves pain conditions and provides better care of Huntington's disease's patients.

A proper positioning of the patient with elements of basal stimulation that support the well-being should be done for a better quality of life with prevention of pain and skin and soft tissue damages in advanced stages. **Basal stimulation is based on the principle of individual own sensations** (favorite music, tastes, massage) that give rise to positive emotions.

Figure 21: Practice examples for patients while lying down

A: Arm Lifts

Start position: Lie flat on your back with your hands by your side.
Exercise: Stretch your arms overhead and hold briefly, then return to the starting position. Repeat several times.

B: Front Punches (Boxing)

Start position: Lie flat on your back with your shoulders relaxed, elbows bent, and hands on your chest. Then make fists with your hands and remain relaxed. Exercise: Punch one arm forwards along the line of your shoulder until your arm is straight, hold briefly, and then return back to the starting position and change sides. Repeat several times.

C: Spinal Rotation

Start position: Lie on your back, with your knees bent, feet flat, and arms slightly spread. Exercise: Rotate your knees to the left. Hold for 30 seconds. Repeat several times. Alternate sides.

D: Knee Extension

Start position: Lie on your back with your knees bent.
Exercise: Straighten your leg at the knee, then return to the starting position. Repeat several times. Alternate sides.

E: The Bridge

Start position: Lie on your back with your hands by your side, your knees bent and feet flat on the floor. Exercise: Tighten your abdominal muscles followed by your buttock muscles. Lift your hips, creating a straight line from your knees to your shoulders. Repeat several times.

F: Air Cycling (Bike)

Start position: Lie on your back and lift both your legs up.
Exercise: Perform a cycling motion with your legs. Repeat several times

Fig. 22: Practice examples for patients while stitting

A: Arm Side Raises

Start position: Sit up straight, and place your hands on your thighs.
Exercise: Raise your right arm up to the side until your arm is straight, turn your head the same direction, hold briefly, then return to the starting position. Repeat several times. Alternate sides.

B: Elbow cirlces (chicken)

Start position: Sit up straight. Place fingertips on your shoulders and extend your elbows. Exercise: Rotate your elbows forward and backwords in a circular motion. Repeat several times.

C: Arms Swing

Start position: Sit up straight on the edge of your seat.
Exercise: Swing your arm, bringing the right arm up and left arm down, alternately. Repeat several times

D: Front Arm Streches

Start position: Sit up straight. Place your fingertips on your shoulders and extend your elbows.
Exercise: Stretch your arms out in front of you and hold briefly, then return to the starting position. Repeat several times.

E: Side Leg Extension

Start position: Sit up straight. Exercise: Slowly straighten one leg out to the side and touch the floor with your heel. Repeat several times. Alternate sides.

F: Seated Leg Extension

Start position: Sit up straight. Exercise: Slowly straighten one leg out to the front and touch the floor with your heel. Repeat several times. Alternate sides.

Figure 23: Practice examples for patients while standing

A: Toe Heel Stand

Start position: Stand behind a chair. Rest your hands on the back of the chair for support.

Exercise: Go up on your tip toes and come down flat. Balance on both toes and hold shortly. Increase your time as you feel comfortable. In the next step lift your toes up off the floor and balance on both heels. Repeat several times.

B: Partial squats

Start position: Stand up straight with your feet apart. Rest your hands on the back of the chair for support.
Exercise: Slowly do small knee bends. Concentrate on squeezing the muscles in your buttocks while you straighten up again. Repeat several times. Alternate sides.

C: Side leg raises

Start position: Stand up straight with your feet apart. Rest your hands on the back of the chair for support. Exercise: Lift your right leg out to the side and swing it back. Repeat several times. Alternate sides.

D: Leg Swing

Start position: Stand up straight with your feet apart. Rest one hand on the back of the chair for support. Exercise: Swing the outside leg forward and backward, slowly letting the weight of your leg gently stretch the hip. Repeat several times. Alternate sides.

E: Front and Back Lunge

Start position: Stand up straight with your feet apart. Rest one hand on the back of the chair for support. Exercise: Step to the front. Bend one knee slightly, keeping the other leg straight. Lean forward with your body, hold briefly and then step back. Repeat several times. Alternate sides.

F: Shoulder Swinging

Start position: Stand up straight with your feet apart and your hands by your side. Exercise: Swing your arms away from your body to the right and left side. Repeat several times.

G: Side Lunge

Start position: Stand with your feet together. Exercise: Step to the side. Bend one knee, keeping the other leg straight. Lean on the knee, hold for 3 seconds and then return it back to the starting position. Repeat several times. Alternate sides.

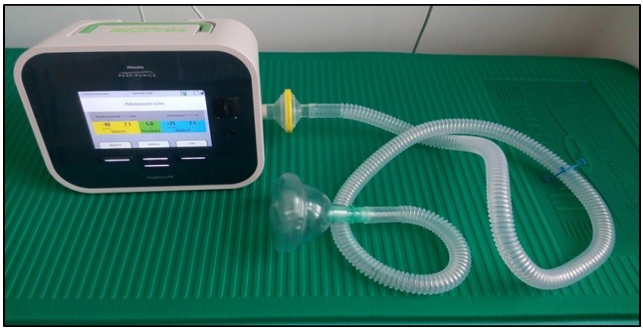
13.5. Prevention of aspiration and respiratory infections

The **regular respiratory muscles training (RMT) should be performed to prevent aspiration and respiratory infections** and improve the function of the respiratory muscles; different devices may be used (figure 24, figure 25, figure 26). It is advisable for an introduction to the respiratory muscle training to consult a physiotherapist or speech language pathologist.

Figure 24: Inhalation and expiratory threshold

Figure 25: Acapella

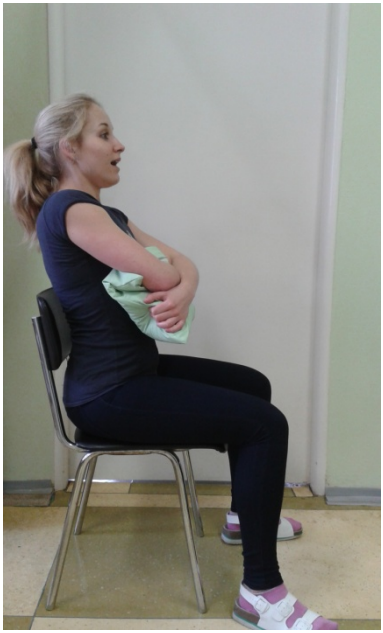
Figure 26: Cough Assist



It is recommended, when coughing in case of respiratory system weakness to stabilize the chest manually (figure 27). The cough effectiveness may be increased by several successive deep breaths to maximum lung volume (maximum insufflation capacity, figure 28).

Figure 27: Manual chest stabilization

Press your chest (e.g. by a pillow), breathe deeply in and then cough intensively



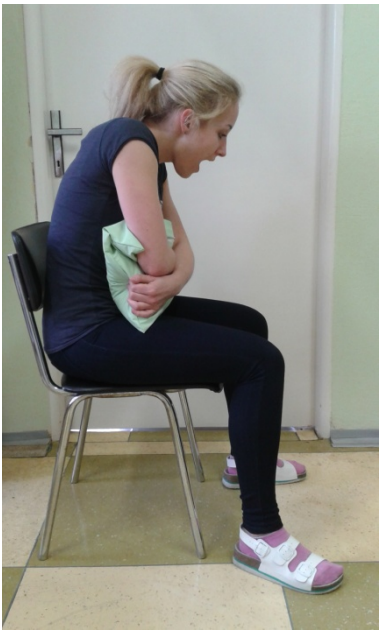


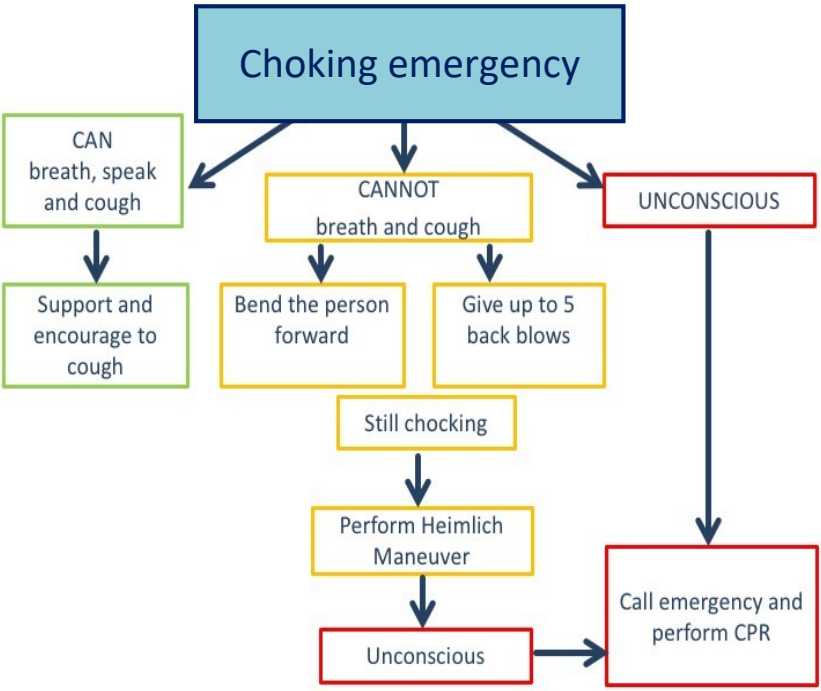
Figure 28: Maximum insufflation capacity

Take three deep breaths of maximum capacity and then cough intensively

13.6. First aid for choking

Choking occurs, when a foreign subject, in case of a Huntington´s Disease patient e.g. a piece of food blocks the airways. In case of choking on a foreign body or substance in the airways, it is recommended to follow the following schedule (figure 29):

Figure 29: First aid scheme for choking



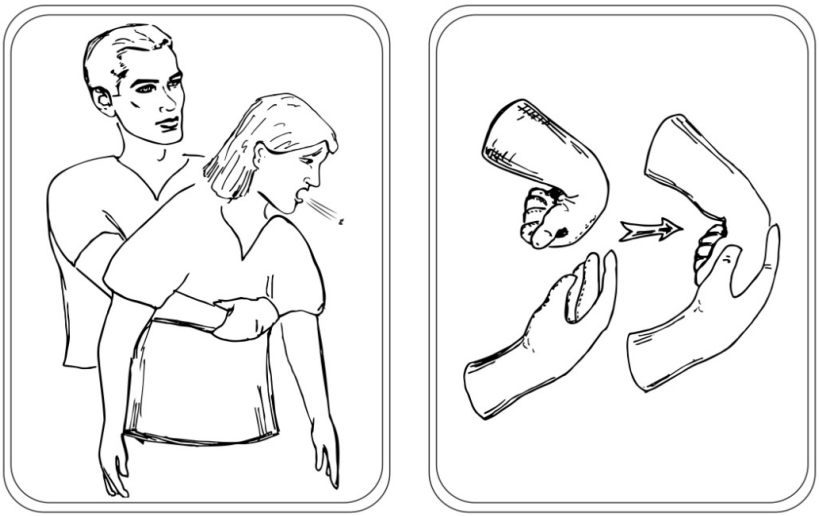
If the affected person is **conscious and able to cough sufficiently**, the rescuer only supports the patient in coughing (natural mechanisms are more effective than first aid support measures).

If person is **conscious, but cannot cough**, talk or breathe, or makes whistling sounds when breathing, the rescuer should stand next to the affected person and lean the affected person slightly forward so that the loose object can fall out of the patient´s mouth. Then deliver five back blows between the shoulder blades with the heel of the hand and check if the airways are released

If the **blockage is still present**, the **Heimlich maneuver should be performed**: The rescuer stands behind the affected person, wraps the hands around the waist and leans the person forward. The rescuer makes a fist from one hand and places it above the navel, with the other hand grasps the fist and presses strongly inward and upward; if needed repeated five times (figure 30).

The rescuer repeats these procedures until he or she removes the airway closure or until the affected person loses consciousness. If the person is unconscious, the rescuer proceeds immediately with urgent resuscitation.

Figure 30: Heimlich's maneuver



Summary

- Physiotherapy should be started immediately after the diagnosis of the disease, do not delay until the first difficulties arise. Early training is important for maximum use of functional reserves.
- Due to the frequent apathy, the motivation to practice is not always 100%, and therefore support of the closest ones (family, friends, care givers) is needed.
- Physical training must be adapted to current physical and psychological abilities.
- Training should be intense and regular.
- Sufficient energy supply is always necessary to combine with physical activity that ensures the building of functional muscle mass.
- As a prevention of aspiration and respiratory infections it is necessary to strengthen the respiratory muscles.

14. Ethical aspects of care in advanced stages of Huntington's Disease

by Jiří Klempíř, Olga Klempířová, Alžbeta Mühlbäck

14.1. Palliative care

Huntington's disease is a chronically progressive disorder. At the moment, we are still missing medication, that could stop, slow down or even cure this disorder (**causal therapy**). Nevertheless, medicine may positively influence a variety of disease symptoms (**symptomatic therapy**) by using different drugs, and contribute to a significant improvement of quality of life of patients and their caregivers. Under these circumstances, in advanced stages we talk about so called **palliative medicine** and its aim is to ensure an acceptable quality of life. The objectives of this therapy are summarized in table 8. Palliative care requires a multidisciplinary approach by physicians, nurses, social workers, psychologists, pastoral counselors, physiotherapists, occupational therapists and nutrition specialists. This team is guided by the needs and wishes of the patient, including the so-called "previously expressed wishes", at the same time, obliged to respect ethical rules and legal standards.

Palliative care may be provided in a hospital, hospice, nursing home or home environment. Unfortunately, society in a lot of European countries is not sufficiently informed about aims and possibilities of palliative medicine, that it does not have to be automatically the care of the patient in his/her terminal stages of the disease. This care is individual and, depending on the type and nature of the illness, can be effectively provided for many months and years, so we should not hesitate to initiate it for a better quality of life.

Tab. 8: Objectives of palliative therapy

Adequate hydration
Adequate nutrition
Proper digestion function
Pain relief
Comfortable breathing
Physical and mental comfort

14.2. Psychological diagnosis and support of the patient and his/her family

Psychological care is a very important part of palliative medicine. Before starting with psychological care of the patient with unfavorable prognosis, it is necessary to find out whether the patient and his or her loved ones have the right of information about the course of the disease and its prognosis. Sometimes unrealistic expectations and doubts may be needed to be cleared and discussed. It is also important, through diagnostic interviewing, observation and sometimes short test methods, to assess the mental state and cognition of the patient, and to focus on the emotional and relationship settings in the family. In a lot of families, there are as well caregivers or family members suffering from anxiety and depression.

The needs of the patient are closely related to the level of cognitive deficits, communication skills, emotional state and disease acceptance, that determine the patient's ability to deal with the illness on the one hand and the maintenance of a good quality of life, including communication with loved ones, on the other hand. When a patient is able to express his/her own needs, we follow these wishes, unless the patient is under the influence of psychotic or depressive thinking with ideas about self-harm or harm to the environment. For apathetic or dementia patients, we try to provide at least the basic needs such as safety, a pleasant atmosphere without fear and to avoid negative stimuli from environment. If the patient is no longer able to express his/her own needs and wishes, we must rely on our observation and experience of the non-verbal expression and basic emotional states - anxiety, sadness, anger, joy, satisfaction, disgust, dislike. Sometimes simple gestures may indicate what the patient wants or rejects. When the patient is expressing a minimum of non-verbal signals, in case of severe apathy or depression, we try to provide only pleasant stimuli as known in the concept of basal stimulation. We may rely on recommendations from family or caregivers who can provide valuable information about the patient´s past habits and favorite activities.

The needs or unrealistic expectations of the family are often based on insufficient understanding of the patient's condition, coping with the gradual loss of a close person. Close relatives and caregivers may really benefit from open communication about the illness and encouraging communication with the patient. Family and caregivers need support when in contact with terminally ill loved ones and mostly at the time of coping with his/her death. They need to be appropriately informed about changes in treatments based on the patient´s current status (medication, nutrition, hydration). Promoting patient communication with family members is important to ensure the patient's sense of belonging and safety. The relatives need regular contact with health care professionals and repeatedly to explain and discuss the patients' health condition.

On the other hand, they should be respected when they decide not to be informed about the course of the disease or treatment.

14.3. Nutrition in palliative care

Nutrition does not affect or cure the underlying disease, but early nutrition intervention improves the quality of life, reduces the incidence of complications (e.g., sores, infections) and **increases the effectiveness and tolerance of other therapies**. From a psychological point of view, it is encouraging if the patient is able to eat. It is important to know that the administration of the indicated nutrition does not prolong the suffering of the affected person.

14.4. Nutrition in the terminal stages of Huntington's Disease

In the terminal stage of the disease, we are choosing another approach. This is the most difficult phase for all involved (patient, family, caregivers, healthcare professionals), and there is no universal procedure for how to handle it. It is very difficult to state, when the **terminal stage of the disease** begins, because the course of Huntington's disease is a continuous process, but the patient´s condition may change unexpectedly and quickly. The following information should help you and guide you through this situation.

In terminal stages, the main effort is to ensure the best acceptable quality of life (tab. 1), but we are no longer prolonging the condition of the terminally ill patient by administration of specific treatment, such as antibiotics, artificial pulmonary ventilation, dialysis, artificial nutrition and medication, which under these circumstances may no longer contribute to an improvement of the patient´s condition (chapters 8, 10 and 11).

The goal of the nutritional therapy at the terminal stage is to provide a nutritional intake on a natural basis. We consider the artificial nutrition and hydration to be treatment intervention, that delivers nutrients and fluids to the body, the patient is not experiencing the same feeling like eating and swallowing on his/her own. Therefore, the artificial nutrition (chapter 8) at the terminal stage of disease may be terminated to overcome complications and unpleasant feelings of the patient, such as nausea, bloating or diarrhea. The introduction of percutaneous gastrostomy at the terminal stage is unethical. It has to be considered, that the placement procedure requires anesthesia, there is also a risk of infection and other side effects and complications. The terminal condition does not require full nutrition and continued reduction, and discontinuation of artificial nutrition does not lead to feelings of hunger and thirst.

Nutrition may even worsen the patient´s condition and shows no potential benefits. The terminally ill patient does not benefit from nutrition as mentioned above. Studies

show that the majority of dying patients are not experiencing hunger, the patient should never be forced to eat. The patient has the right to choose, what and when he/she is eating. At this point the family and caregivers need support and a lot of explanations about the effectiveness or lack of artificial nutrition and feeding to better understand the terminal process and the condition of the terminally ill patient.

14.5. Hydration in terminal stages of Huntington Disease

In the same way, as hunger and interest in food is decreasing in the terminal stage, the terminally ill patient is not experiencing the feeling of thirst and is drinking less. We may try to encourage an affected person to frequently drink small amounts of fluid, but we have to respect at the same time, the right of the person to reject it. Generally, the affected person needs a reduced amount of fluids, not more than half of the amount that he was used to. The higher the fluid intake, the larger problems and complications, such as nausea, edema and worsening of heart and kidney functions. The feeding tube may be used for hydration, although the intravenous and subcutaneous infusions should be given only exceptionally.

A small amount of alcohol or beer, as wished by the patient may improve the appetite. Chewing gum may increase the production of saliva and reduce feelings of nausea. Sour candies may stimulate saliva production. A lot of water is contained in soups, sauces, fruits, vegetables and ice cream that should be considered for hydration. Water or flavored ice cubes reduce the dry feeling in the mouth.

Summary

- Nutrition does not affect or cure the underlying disease, but early nutrition intervention improves the quality of life, reduces the incidence of complications (e.g., sores, infections) and increases the effectiveness and tolerance of other therapies.
- Early and properly indicated nutrition and hydration improves the physical condition and does not prolong suffering.
- The inability to eat or lack of interest in food and drink is part of the terminal process (dying).
- We always respect the patient's eating habits and wishes.
- Not only an affected person in the terminal phase, but both relatives and caregivers need professional help.
- If the caregiver or family members have doubts about the patient management in the terminal stage, they should consult an expert on palliative care.
- If the patient´s opinion differs from a caregiver´s one, it may be useful to consult a physician or expert on palliative care.
- All open questions have to be answered.

15. Organisation of nutrition care

by Jiří Klempíř, Alžbeta Mühlbäck

At present, we are overwhelmed by advertisements promoting healthy nutrition and different nutritional programs and coaching. Unfortunately, those services are sometimes of poor and insufficient quality and are not suitable as a consultation or replacement of the nutritional specialist. To guarantee quality nutrition consultation by nutrition professionals such as dietitians, certified nutritionists (clinical nutritionist), appropriate evidence of professional experience and registered according to the regulations of different countries, is necessary. There is as well the possibility to consult a consulting physician or physician nutrition specialist.

Physician Nutrition Specialist (PNS) is a physician who has obtained training in nutrition and its management in multidisciplinary patient care.

Dietitians or Certified Nutritionists after completing higher education can act independently in the field of diagnostics, prevention and treatment of nutrition disorders. They are like any medical practitioner obliged to follow rational and proven procedures and guidelines. The nutrition assistant (NA) is performing the work of a health care professional under professional supervision and in consultation with a dietitian or certified nutritionist. Nutrition assistants may be working at different institutions such as schools, hospitals, clinics, nursing homes, etc.

It is important to know that a number of professions such as diet and nutrition experts, health coaches and nutritional therapists do not comply with the high educational standards as dietitians or certified nutritionists and are not experts on nutrition disorders and clinical nutrition.

Summary

- The best way of management of nutrition disorder is to consult professionals and experts in this field such as physician nutrition specialists, dietitians or certified nutritionists.

Authors and Affiliations

Assoc. Prof. Jiri Klempir, MD, PhD

Neurologist, physician nutrition specialist

Department of Neurology and Centre of Clinical Neuroscience, First Faculty of Medicine and General University Hospital, Charles University in Prague, Prague, Czech Republic

Institute of Anatomy, First Faculty of Medicine, Charles University in Prague, Prague, Czech Republic

Jiří Klempíř is a neurologist, working at the Movement Disorders Centre in Prague which has over 25 years expertise in Huntington's Disease.

His main interests are focused on neurodegenerative disorders with a special emphasis on movement disorders like Parkinson's and Huntington's disease. The multidisciplinary team at the Movement Disorder Center provides comprehensive neurological, psychiatric, psychological and physiotherapeutic care and as well offers regular ambulatory consultations for HD patients and their relatives including genetic counselling.

Jiří Klempíř has an outstanding experience in nutritional management in HD and is closely collaborating with dietitians, speech and swallowing therapists, focusing on early diagnostic of dysphagia, prevention and treatment of malnutrition. He is supervising pregradual and postgradual students in the field of Neurosciences and actively participates in clinical and basic research.

Centre collaborates with Czech Huntington Association, European Huntington Association, European Huntington´s Disease Network and European Reference Network on Rare Neurological Diseases.

Alžbeta Mühlbäck, MD

Neuropsychiatrist

Huntington Center South, kbo Isar Amper Klinikum – Klinik Taufkirchen, Taufkirchen, Germany

Department of Neurology and Centre of Clinical Neuroscience, First Faculty of Medicine and General University Hospital, Charles University in Prague, Prague, Czech Republic

Alžbeta Mühlbäck is the senior consultant for neuro-psychiatry at the Huntington Center South in Taufkirchen, Germany.

She was born in Slovakia, former Czechoslovakia and graduated in 2004 from the Medical School of Olomouc University, Czech Republic, afterwards being active as a physician in Austria and Germany.

Since 2010 Alžbeta is working in Taufkirchen, initially in the department of general psychiatry, soon after in the neuro-psychiatric ward for Huntington's Disease, where she practices as a senior consultant.

In this function she is taking care of the HD department and as well offers regular ambulatory consultations for HD patients and their relatives including genetic counseling.

Alžbeta is closely collaborating with EHA (European Huntington Association) and together with EHA she planned and launched the online service for the HD community: 'Ask The Doctor' on the EHA website, where the HD community can ask any kind of question in privacy.

Her years-long expertise in psychiatry, neurology and genetics is perfectly well adapted to the multi-faceted face of HD. With regards to HD research activities Alžbeta is collaborating with Charles University in Prague, Czech Republic. Alžbeta is as well a member of the EHDN Working Group for Genetic Testing and Counseling. She acquired research experience from several clinical trials (e.g. Amaryllis trial, DBS -HD) as well as from the Enroll-HD study.

Mgr. Eva Baborová

Speech and swallowing therapist

Department of Neurology and Centre of Clinical Neuroscience, First Faculty of Medicine and General University Hospital, Charles University in Prague, Prague, Czech Republic

Bc. Martina Dvořáková

Nurse

General University Hospital in Prague, 4th Department of Internal Medicine, Prague, Czech Republic

Bc. Petra Havrlíková

Nurse

General University Hospital in Prague, 4th Department of Internal Medicine, Prague, Czech Republic

PhDr. Olga Klempířová, PhD

Clinical psychologist, neuropsychologist

Department of Neurology and Centre of Clinical Neuroscience, First Faculty of Medicine and General University Hospital, Charles University in Prague, Prague, Czech Republic

Psychiatric Hospital Bohnice, Prague, Czech Republic

MPT Romana Konvalinková

Physiotherapist

Department of Neurology and Centre of Clinical Neuroscience, First Faculty of Medicine and General University Hospital, Charles University in Prague, Prague, Czech Republic

Lucie Růžičková, Bc.

Head dietitian

General University Hospital in Prague, Department of Dietitians, Prague, Czech Republic

This publication summarizes all relevant and practical information about the nutritional management in Huntington Disease. It covers all the topics from the early detection of nutritional problems, explaining their causes and consequences, to the management of dysphagia to the nutrition in advanced stages of Huntington Disease in plain language. Moreover, it provides practical recommendations and solutions. The publication is in form and content primarily addressed to patients, their families and caregivers or medical staff to receive compact information on the issue of nutritional disorders within Huntington Disease.

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