Nutritional Support in Neurological Diseases

Module 25.4
Nutritional Support in Chronic Neurological Diseases

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Learning Objectives

- To know about nutritional intervention in patients suffering from multiple sclerosis or Parkinson’s disease;
- To know the problems of clinical decision making in the timing/safety/efficacy of percutaneous endoscopic gastrostomy (PEG) tube feeding in amyotrophic lateral sclerosis (ALS) patients;
- To understand the clinical challenges of establishing and maintaining tube feeding in neurological patients with advanced dementia;
- To discuss the indications for gastrostomy in neurological patients with advanced dementia;
- To know the ethical, legal, and moral implications of nutritional support in patients with neurodegenerative disorders with progressive dementia.

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Key Messages

- Patients with chronic neurological diseases are at risk of malnutrition, with dysphagia the most frequent concern in terms of evaluation and treatment;
- Amyotrophic lateral sclerosis patients generate the most frequent dilemmas concerning timing/efficacy and security of PEG placement;
- There are no robust data available to support tube feeding over oral feeding in advanced terminal dementia;
- Patients with advanced dementia are not usually considered appropriate candidates for artificial feeding via PEG tubes;
- Legally and ethically, specialized nutritional support should be considered to be a medical therapy;
- Advanced directives help to resolve conflicts over desired treatments.
1. Introduction

Neurodegenerative diseases are a heterogeneous group of disorders that are characterized by the progressive degeneration of the structure and function of the central nervous system (CNS) or peripheral nervous system. The CNS acts as a regulator of nutrient intake by internal signalling mechanisms, which become disrupted in CNS injury. Chronic, degenerative diseases of the brain can also cause significant complications affecting the nutritional status, which develop gradually as the disease progresses. Residual neurological function often changes over time, generating different nutritional concerns depending on the stage of CNS injury (1). Dysphagia is the most frequent concern in terms of evaluation and treatment (2). Nutritional support should be considered as a valuable adjunctive therapy in chronic neurological patients with malnutrition or risk of malnutrition. The objectives of the nutritional support can vary widely among diseases. In multiple sclerosis or Parkinson’s disease the objective can be to cover all the nutritional requirements of the patient. In chronic neurological diseases with advanced dementia like Alzheimer’s disease, the objective may be comfort feeding and reinforcement of quality of life. Nevertheless, a great concern exists about the possibility of preventing or delaying the progression of chronic neurological disease with adequate nutritional support (for more details, see the LLL Module 20.4).

2. Nutritional Support in Chronic Neurological Conditions

2.1 Multiple Sclerosis

Multiple sclerosis (MS) is the most common degenerative auto-immune disorder of the CNS. In the acute phase of MS, T-lymphocytes attack oligodendrocytes and this leads to inflammation and scarring of the myelin sheaths. The conduction of nerve impulses along the axon of the neurone may be affected during an acute inflammatory phase (relapse), but tends to improve with healing during the remission phase. Over time, relapses cause extensive damage and scarring of the myelin sheath with progressive loss of neuronal function. The cause of MS is unknown, but research suggests that genetic, immunological and environmental factors, such as a common virus, may all be involved in a complex aetiology (3).

Symptoms of MS vary widely between individuals, depending on the site of scarring, and can affect brain, peripheral nerves and spinal cord. There is no direct evidence that nutrition is involved either in the aetiology of MS or in the rate of disease progression. Epidemiological studies have shown that MS is more prevalent in countries with a high intake of saturated fat, and lower in countries with high intakes of polyunsaturated fat. However, intervention studies using modifications in fat intake as well as modulation of n-6 and n-3 fatty acids to try and reduce the rate and severity of relapse in MS have shown conflicting results (4).

Antioxidants to inhibit oxidation of essential fatty acids by free radicals in the membrane phospholipids of the myelin sheath have also been proposed. Although there is a growing interest in dietary antioxidants and disease activity in MS, there is no scientific evidence that any form of dietary intervention is effective in slowing the rate or severity of this disease. Although initially a favourable relationship was postulated, a meta-analysis has been published highlighting the lack of robust evidence on the use of vitamin D and multiple sclerosis relapses (5). Patients with MS are at risk of malnutrition as the degree of disability progresses. For more details, consult module 25.1.

Nutrition management

Overweight and obesity can also occur initially due to reduced mobility, corticosteroids, antidepressants drugs, and unhealthy dietary habits (use of fatty foods, sugary drinks,
etc.) In these cases, a review of dietary habits provides the opportunity to reinforce healthy eating advice (4).

With increasing disability, weight loss, malnutrition and cachexia are frequent. Although the incidence of malnutrition in MS patients has not been determined, it can adversely affect organ function, muscle strength and immune function, thereby worsening the symptoms of MS (6). The possible causes of malnutrition are summarized in Table 1. Dietary management of weight loss depends upon identification of the source of the problem.

Table 1
Aetiology of weight loss and related dietary interventions in patients with multiple sclerosis

<table>
<thead>
<tr>
<th>CAUSE</th>
<th>INTERVENTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reduced mobility and fatigue</td>
<td>Assistance with shopping and cooking</td>
</tr>
<tr>
<td>Difficulty with shopping and cooking</td>
<td></td>
</tr>
<tr>
<td>Physical difficulty manipulating food or fluid to mouth (tremor or postural difficulty)</td>
<td>Finger foods, lightweight utensils, special feeding utensils</td>
</tr>
<tr>
<td>Poor sight</td>
<td>Assistance with shopping and cooking, and with eating and drinking</td>
</tr>
<tr>
<td>Quickly fatigued when eating, so small meals and few between-meal snacks</td>
<td>Small frequent meals and between-meal snacks. Fortify foods. Dietary supplements</td>
</tr>
<tr>
<td>Poor appetite</td>
<td>Consider side-effects of drug treatment. Small frequent meals and snacks. Fortify foods. Dietary supplements</td>
</tr>
<tr>
<td>Reduced cognition</td>
<td>Extra assistance</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>Modified textured diets / EN</td>
</tr>
<tr>
<td>High dependence on reduced-fat convenience meals</td>
<td>Liberal use of full-fat dairy products, increase snacks rich in vegetable oils</td>
</tr>
<tr>
<td>Adverse effect of drugs</td>
<td>Evaluate if all are needed</td>
</tr>
</tbody>
</table>

Dysphagia is a symptom of chronic progressive MS. The reported incidence ranges between 3 and 43% depending on the method used to evaluate the swallowing difficulty (7-10). It is often accompanied by speech difficulties. Features of dysphagia include coughing and choking during meals, frequent chest infections and weight loss. The evaluation of the ability to swallow may be clinical initially but this should be followed by video fluoroscopy or fibreoptic endoscopic evaluation of swallowing. The type of nutritional intervention depends on the degree of difficulty, and ranges between modified consistency diets and exclusive artificial nutrition if the risk of aspiration is very high.

If weight loss progresses despite oral intervention or if the dysphagia cannot be treated safely by dietary modifications, enteral nutrition via a PEG should be considered. Enteral tube feeding can improve nutritional status, reduce the risk of aspiration pneumonia, reduce the risk of pressure sores, and maintain fluid balance as well as allowing some continuing oral intake. However, before embarking on treatment, the pros and cons of artificial nutrition should be discussed fully with the patient and family.

2.2 Parkinson’s Disease

Parkinson’s disease (PD) is a chronic, progressive neurodegenerative disorder resulting from dopamine depletion in the brain. The main symptoms include tremor, muscular rigidity, bradykinesia and postural instability. As PD progresses, a variety of other symptoms emerge, including dysphagia, dysarthria, impaired gastrointestinal motility, fatigue, depression and cognitive impairment. Drug therapy is essential to control symptoms and to maintain mobility in PD, and acts by replacing or mimicking dopamine in the brain.
Patients with PD are at increased risk of malnutrition and weight loss (11), and nutritional status should be monitored routinely throughout the natural history of the disease (12, 13) (see module 25.1).

Dysphagia in PD usually occurs in the advanced phases of the disease, although sometimes it is present from the onset. Functional alterations in oropharyngeal and oesophageal motility can be present in about 60-80% of patients, but may be asymptomatic. Impaired gastric emptying is commonly reported in patients with PD at both early and advanced stages. It seems to be related to severe motor function impairment and motor fluctuations, typical of disease progression. It can be treated with domperidone. Malnutrition in PD patients is probably under-reported, and can be found in about 15% in community-dwelling patients with PD, and another 24% of patients are at medium or high risk of malnutrition. Several predictors of malnutrition have been found: older age at diagnosis, higher levodopa equivalent daily dose / body weight, greater UPDRS score, anxiety and depression, and living alone (13).

Amino acids compete with levodopa for transport across the blood-brain barrier, and it was therefore postulated that low protein diets might improve drug uptake and enhance mobility in patients taking levodopa. However, dietary protein restriction seems to be helpful in only a small number of patients who have severe medication-induced fluctuations in mobility, and is not recommended for continuous use. Rather than restricting protein, patients should be advised to redistribute protein throughout the day and to avoid taking high protein foods at the same time as their levodopa. With the introduction of controlled release preparations of levodopa the proportion of patients with PD who might benefit from protein restriction has declined. In severe cases, duodenal infusion of levodopa (duodopa) can be explored (14, 15).

In summary, in PD our primary nutrition intervention will be focused on healthy dietary intake and minimising adverse drug-nutrient interactions to avoid weight loss and to manage malnutrition and dysphagia.

2.3 Neuromuscular Diseases (Amyotrophic Lateral Sclerosis)

Malnutrition is a common problem among patients with neuromuscular diseases, particularly in those with motor neurone disease. Amyotrophic lateral sclerosis (ALS) is a prototype model of neurodegenerative disease in this respect, characterized by the progressive loss of motor neurones, resulting in the progressive wasting of skeletal muscles, including the respiratory muscles.

The recognition of nutrition as an independent, prognostic factor for survival and disease complications in ALS has highlighted the importance of the nutritional management of these patients (16,17). Patients who have lost weight at diagnosis have a doubled risk of death, and malnourished patients show a 7.7-fold increase in mortality (17).

The aetiology of malnutrition in ALS patients is multifactorial (16):
- The bulbar degeneration of neurones manifests as difficulties with mastication, impaired oral transit, prolonged meal times and dysphagia.
- Anorexia is frequent and commonly attributed to psychological distress, depression and polypharmacy.
- The weakening of abdominal and pelvic muscles, limited physical activity, self restriction of fluids and lower-fibre diets can result in constipation, which in turn can exacerbate the anorexia.

Despite the reduction in lean body mass, ALS patients have increased energy needs, due to increased respiratory efforts, pulmonary infections and other factors yet not established (18). Among patients suffering from respiratory failure, non-invasive home ventilation is commonly practiced in most major European centres, and this may mean that energy requirements need reassessment to avoid weight loss and net catabolism, and also because of the effects of carbohydrate and protein intake on metabolic rate and the demands for increased gas exchange.

Malnutrition exacerbates the disease related breakdown and atrophy of skeletal and respiratory muscles, and impairs the immune system, predisposing to infection, a common cause of death in ALS patients (17).
The key issues for nutritional management in amyotrophic lateral sclerosis include (19,20):
- Energy supplementation
- Diagnosis/treatment of dysphagia

Energy supplementation

Recently, NICE Guidelines on motor neurone disease have been published (21), and some recommendations are summarized about oral nutritional support in ALS.

1. At diagnosis and regularly, we need to assess the person’s weight, personal diet, nutritional and fluid intake, hydration, oral health, feeding, drinking and swallowing, and offer support, advice and interventions as needed. To design nutritional intervention, we need to assess appetite and thirst, gastrointestinal symptoms, such as nausea or constipation, and evaluate causes of reduced oral intake (for example, swallowing difficulties, limb weakness or the possibility of low mood or depression causing loss of appetite.

2. Assess the person’s ability to eat and drink by taking into account:
   a. the need for eating and drinking aids and altered utensils to help them take food from the plate to their mouth.
   b. the need for help with food and drink preparation.
   c. advice and aids for positioning, seating and posture while eating and drinking.
   d. dealing with social situations (for example, eating out).

3. Arrange for a clinical swallowing assessment if swallowing problems are suspected.

4. Assess and manage factors that may contribute to problems with swallowing, such as: positioning, seating, the need to modify food and drink consistency and palatability, respiratory symptoms and risk of aspiration and/or choking, and fear of choking and psychological considerations (for example, wanting to eat and drink without assistance in social situations).

When considering diet, although the available evidence showed no benefit of supplements in terms of survival, food supplementation may be useful for some people and should be considered on an individual basis following a nutritional assessment. The NICE Guidelines identified that it was preferable to increase calorie intake by enriching food instead of providing food supplements.

Diagnosis and treatment of dysphagia

Dysphagia in ALS patients is highly prevalent, and can be the initial symptom in bulbar onset ALS. In advanced forms of the disease, the prevalence of dysphagia can be near 100%. For more details about diagnosis of dysphagia, refer to module 25.1.

Gastrostomy

Mortality in ALS patients undergoing PEG tube insertion is strongly correlated with respiratory function. Practice guidelines from the American Academy of Neurology recommend that the procedure should be performed when a patient’s forced vital capacity is still greater than 50% of the predicted value (for more information about the placement of PEG tube see module 25.2). Recent reports show, however, that many PEGs have been placed in patients who have not met this criterion (22), and several studies have proved that lower vital capacity at the time of PEG insertion is not associated with poor outcome.
when non-invasive positive pressure ventilation is used or if the gastrostomy procedure is carried out radiologically. Patients may benefit from an open discussion of the benefits and hazards of PEG early in the course of the disease and before swallowing problems have developed. Changes in nutritional status and the identification of unintentional weight loss or dysphagia are the best guides to the timing of PEG placement so as to maximize quality of life. A Cochrane systematic review of the literature showed that there is no evidence for efficacy of enteral feeding in ALS patients in terms of mortality, although there are possible advantages in other respects: of nutritional status (only one prospective study) and quality of life (understudied) (20). Probably, these results are a reflection of the lack of sufficient randomized controlled trials due in turn to ethical difficulties and problems of study design. The American Academy of Neurology, Quality Standards Subcommittee, has published an evidence-based review with regard to drugs, nutrients and respiratory therapies for patients with ALS (22). It was concluded that PEG feeding should be considered to stabilize weight and to prolong survival in patients with ALS (Level B). A nutrition management algorithm is proposed *(Fig. 1).*

![Fig. 1 Nutrition management algorithm for patients with ALS (10)](image)

The NICE Guidelines 2016 for motor neurone disease (MND) / ALS (21) recommend:

- to discuss gastrostomy at an early stage, and at regular intervals as MND progresses, taking into account the person’s preferences and issues, such as ability to swallow, weight loss, respiratory function, effort of feeding and drinking and risk of choking. The early placement of a gastrostomy can help to ease a person’s discomfort during the progression of MND by preventing malnutrition through enteral administration of fluids,
food and medication. This may prolong survival. Be aware that some people will not want to have a gastrostomy.

- Explain the benefits of early placement of a gastrostomy, and the possible risks of a late gastrostomy (for example, low critical body mass, respiratory complications, risk of dehydration, different methods of insertion, and a higher risk of mortality and procedural complications).
- If a person is referred for a gastrostomy, it should take place without unnecessary delay.
- Pay particular attention to the nutritional and hydration needs of people with MND who have frontotemporal dementia and who lack mental capacity. The multidisciplinary team assessment should include the support they need from carers, and their ability to understand the risks of swallowing difficulties.
- Before a decision is made on the use of gastrostomy for a person with MND who has frontotemporal dementia, the neurologist from the multidisciplinary team should assess the following:
  - The person’s ability to make decisions and to give consent.
  - The severity of frontotemporal dementia and cognitive problems.
  - Whether the person is likely to accept and cope with treatment.
- Discuss with the person’s family members and/or carers (as appropriate; with the person’s consent if they have the ability to give it).

3. Enteral Feeding Considerations in Patients with Chronic Neurological Disorders

Several questions need to be addressed when considering the enteral nutrition of patients with chronic neurological diseases (23):

1. Patient’s level of consciousness
2. Can the patient protect the airway from episodes of gastro-oesophageal reflux?
3. What is the patient’s ability to swallow liquids, thickened liquids, and soft and solid foods?
4. Is the patient at high risk of aspirating oropharyngeal secretions?
5. Will the need for enteral access be short- or long-term?
6. What are the nutrient requirements of patients with specific conditions (such as ALS)?
7. Is there a difference between gastric and jejunal feeding in this population?
8. What are the patient’s and family’s desires with regard to nutritional treatment?

All these questions can help us to decide the most appropriate form of nutritional support in chronic neurological patients and how to prevent the most serious complications such as bronchial aspiration. The causes are summarised in (Fig. 2).
4. Tube Feeding / PEG Placement in Neurodegenerative Disorders

Among the neurodegenerative disorders with progressive dementia, epidemiological data suggest that nutrition can be involved in the aetiopathogenesis of Alzheimer’s disease (AD), and thus that nutritional intervention can potentially be used in the prevention of the disease. The relative contributions of macro- and micronutrients are not known (24). The most recent studies suggest benefit of some nutritional compounds to attenuate the neurodegenerative process (e.g. antioxidants and free radical scavengers, omega-3 fatty acids, Mediterranean diet) (25). This could be of enormous interest in the initial phases of the disease.

On the other hand, deficiencies in zinc and vitamins E and C are associated with cognitive dysfunction in AD patients, but replacement or supplementation does not lead to improved neurological outcome (26).

Involuntary weight loss is common and may be one of the initial symptoms as AD progresses. The symptoms associated with AD make self-feeding progressively more difficult; therefore, malnutrition is common.

Nutritional status should be assessed at the time of diagnosis, and reassessed during follow-up. Weight loss or anorexia must alert the caregivers and warrants full nutritional assessment. We need to plan initial nutritional intervention if weight loss is ≥ 5% in 3-6 months.

The disturbances in feeding behaviour observed in AD are dominated by anorexia, which may develop from the onset of the disease. As the disease progresses, feeding behaviour becomes more profoundly disturbed, and can be measured with the Blandford Aversive Feeding Behaviour Inventory. This scale differentiates four types of disturbances:

- Selective behaviours (refusal to eat a wide range of foods). These disturbances require changes in diet.
- Active behaviours (resistance to feeding, with defensive reflexes)
- Oral dyspraxia and attention disorders
- Oropharyngeal dysphagia (loss of muscular coordination during mastication and swallowing, that can lead to repeated problems with swallowing and choking)

In the clinical course of advanced dementia, the prevalence of eating disorders is very high, as is the overall mortality related to eating problems. Recently, the ESPEN Guidelines on nutrition in dementia have been published (27). In the guidelines, the different nutritional problems arising from different disease stages are identified (Table 2).

Table 2
Nutritional problems along the stages of dementia

<table>
<thead>
<tr>
<th>Nutritional problem</th>
<th>Stage of dementia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Olfactory and taste dysfunction</td>
<td>Preclinical and early stages</td>
</tr>
<tr>
<td>Attention deficit</td>
<td>Mild to moderate</td>
</tr>
<tr>
<td>Executive function deficits (shopping, preparing food)</td>
<td>Mild to moderate</td>
</tr>
<tr>
<td>Impaired decision-making ability (slowdown in food choice, reduced intake)</td>
<td>Mild to moderate</td>
</tr>
<tr>
<td>Dyspraxia (coordination disorders, loss of eating skills)</td>
<td>Moderate to severe</td>
</tr>
<tr>
<td>Agnosia (Loss of ability to recognize objects or comprehend the meaning of objects, which means that food may not be distinguished from non-food and that eating utensils are not recognized as what they are)</td>
<td>Moderate to severe</td>
</tr>
<tr>
<td>Behavioural problems</td>
<td>Moderate to severe</td>
</tr>
<tr>
<td>Oropharyngeal dysphagia</td>
<td>Moderate to severe</td>
</tr>
<tr>
<td>Refusal to eat</td>
<td>Severe</td>
</tr>
</tbody>
</table>

Practical guidelines for the diagnosis and management of weight loss in AD (28)

- A varied and balanced diet and daily physical activity should be recommended for every AD patient to help prevent weight loss.
- Search for reversible medical or socioenvironmental causes of weight loss (including drugs, and especially acetyl cholinesterase inhibitors in the first weeks after the onset of treatment).
- Health-care professionals and family caregivers should follow training centred on the detection of nutritional risk and on the food/diet of these patients.
- Planning meals (enriched, fractioned, finger foods).
- Making the patient drink regularly during the day. Prevention of dehydration is a part of the management of AD patients. Nourishing drinks are particularly recommended.

Oral supplementation is needed when patients don’t meet their nutritional requirements with an enriched diet. However, patients with advanced AD may be unable or unwilling to consume an adequate amount of calories by mouth, or may be at risk from dysphagia. In this situation, which can be very distressing for clinicians and families, tube feeding could be seen as a potential option.

The decision to start tube feeding is one of the most challenging dilemmas. The evidence suggests that it may provide more burden than benefit, and we do not have enough data available to support tube feeding vs oral feeding in advanced dementia in the prevention of aspiration pneumonia, reduction of the risk of pressure sores, reducing the risk of infection, improvement in overall function, or in prolonging survival (29).
Moreover, tube feeding represents a considerable use of health care resources. However, we must consider that decisions may and should change with time and clinical situation. ESPEN guidelines recommend tube feeding for a limited period of time in patients with mild or moderate dementia, to overcome a crisis situation with markedly insufficient oral intake, if low nutritional intake is predominantly caused by a potentially reversible condition. However, they do not recommend tube feeding once patients have terminal dementia (27).

The use of PEG is also a matter of debate. The indications for placement of PEG tubes should be when they are expected to improve quality of life, reduce episodes of aspiration, reduce frequency of pressure sores and promote wound healing. However, their benefits have been questioned in patients with advanced dementia. Some studies have shown a worse prognosis in patients who underwent PEG for advanced dementia, in comparison with other diagnoses that require PEG (30), while other studies have shown no differences in survival in patients with advanced dementia between those with and without PEG. In the literature there is accordingly some controversy about the placement of PEG tubes in advanced dementia. Some studies don’t agree that dementia is a negative risk factor for outcome, but acknowledge poor overall survival (31). Other studies underline the need to design prospective controlled studies (32). Overall, the published data support an individualized but critical and restrictive approach to PEG feeding in elderly demented patients.

The recommendations should be addressed so as to improve oral intake and avoid aspiration, and to maintain patient comfort and optimise intimate patient care. The concept of comfort feeding includes careful hand feeding, with an individualized care plan that prioritizes patient comfort, and avoids the misleading dichotomy of care versus no care (33). The comfort of the patient is of primary importance, even in the setting of weight loss.

Recently, in the ESPEN Guidelines for Nutrition in Dementia (27), several recommendations have published:

<table>
<thead>
<tr>
<th>Recommendation</th>
<th>Grade of evidence</th>
<th>Strength of recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>We recommend screening every person with dementia for malnutrition. In case of positive screening, assessment has to follow. In case of positive assessment, adequate interventions have to follow.</td>
<td>Very Low</td>
<td>Strong</td>
</tr>
<tr>
<td>We recommend close monitoring and documentation of body weight in every person with dementia.</td>
<td>Very Low</td>
<td>Strong</td>
</tr>
<tr>
<td>We recommend provision of meals in a pleasant, homelike atmosphere.</td>
<td>Moderate</td>
<td>Strong</td>
</tr>
<tr>
<td>We recommend provision of adequate food according to individual needs with respect to personal preferences.</td>
<td>Very Low</td>
<td>Strong</td>
</tr>
<tr>
<td>We recommend to encourage adequate food intake and to provide adequate support.</td>
<td>Very Low</td>
<td>Strong</td>
</tr>
<tr>
<td>We do not recommend the systematic use of appetite stimulants.</td>
<td>Very Low</td>
<td>Strong</td>
</tr>
<tr>
<td>We recommend educating caregivers to ensure basic knowledge on nutritional problems related to dementia and possible strategies to intervene.</td>
<td>Low</td>
<td>Strong</td>
</tr>
<tr>
<td>We recommend elimination of potential causes of malnutrition as far as possible.</td>
<td>Very Low</td>
<td>Strong</td>
</tr>
</tbody>
</table>
We recommend avoiding dietary restrictions. | Very Low | Strong
---|---|---
We do not recommend the use of omega-3-fatty acid, vitamin B1, vitamin B6, vitamin B12/folic acid, vitamin E, selenium, copper nor vitamin D supplementation in persons with dementia for correction of cognitive impairment or prevention of further cognitive decline. | Different grades | Strong
We recommend the use of ONS to improve nutritional status. | High | Strong
We do not recommend the use of ONS in persons with dementia to correct cognitive impairment or prevent further cognitive decline. | Moderate | Strong
We do not recommend the systematic use of special medical foods for persons with dementia to correct cognitive impairment or prevent further cognitive decline. | Low | Strong
We do not recommend any other nutritional product for persons with dementia to correct cognitive impairment or prevent further cognitive decline. | Very Low | Strong
We recommend that each decision for or against artificial nutrition and hydration for patients with dementia is made on an individual basis with respect to general prognosis and patients’ preferences. | Very Low | Strong
We suggest tube feeding for a limited period of time in patients with mild or moderate dementia, to overcome a crisis situation with markedly insufficient oral intake, if low nutritional intake is predominantly caused by a potentially reversible condition. | Very Low | Weak
We recommend against the initiation of tube feeding in patients with severe dementia. | High | Strong
We suggest parenteral nutrition as an alternative if there is an indication for artificial nutrition, as described in recommendation 16, but tube feeding is contraindicated or not tolerated. | Very Low | Weak
We suggest parenteral fluids for a limited period of time in periods of insufficient fluid intake to overcome a crisis situation. | Very Low | Weak
We recommend against the use of artificial nutrition (enteral nutrition, parenteral nutrition and parenteral fluids) in the terminal phase of life. | Very Low | Strong

5. Ethical Considerations around Tube Feeding in Patients with Neurodegenerative Disorders with Advanced Dementia

Consent obtained from the patient, if he or she is competent, is necessary before commencing tube feeding. But if the patient is not competent, the consent must be obtained from the individual with legal power of attorney. In some jurisdictions this will be the next of kin, but it may also be the attending physician. In all circumstances when the patient is not competent, the physician has to act in the patient’s best interest, and the effects on quality of life and functional status may be the most relevant outcomes to be considered (34).
Even if no advanced directive is available, we need to give priority to the previously expressed wishes of the patient, and be guided by what it is believed the patient would choose if they were competent, always following the patient’s best interest.

If there is a disagreement, we can consider approaching a Clinical Ethical Committee for advice. The establishment of such a committee in a growing number of healthcare institutions will facilitate this process.

Legally and ethically, Specialized Nutritional Support (SNS) should be considered a medical therapy. For this reason, adult patients or their legally authorized surrogates have the right to accept or to refuse SNS. The benefits and burdens of SNS should be considered before offering this therapy, and institutions should develop clear policies regarding withdrawal from or withholding SNS.

Recently, the ESPEN Guidelines on ethical aspects on artificial nutrition and hydration (35) has reached strong consensus on the following recommendations:

- The will of the adult patient who is capable of providing consent and making judgments has to be respected in any case.
- The patient or his or her authorized representative should be informed about the nature, significance and scope of the measure, including potential complications and risks.
- In the case that a patient is unable to give consent and make judgments, the representative (authorized according to different rules depending on the country’s law and practice) takes the decision. The representative has to implement the presumed will of the patient. If the representatives' decision is delayed, the physician should start artificial nutrition according to evidence based medical indications.
- Quality of life must always be taken into account in any type of medical treatment including artificial nutrition.

6. Summary

Chronic neurological patients are a heterogeneous group including those in the chronic phase of an acute injury (acute stroke, brain trauma, and spinal cord injury) and also patients with chronic neurodegenerative diseases, some of them with a progressive and fatal course. Malnutrition is a risk factor for increased morbidity and mortality, and nutritional support plays an important role in the management of all of these diseases. Dysphagia is the major concern that should be identified and treated as early as possible. If dysphagia is severe or cannot be managed with dietary counselling or modified consistency food, enteral tube feeding should be offered to the patient.

In neurodegenerative disorders such as multiple sclerosis or amyotrophic lateral sclerosis, the most important concerns will be the management of dysphagia and the decision to start enteral nutrition via a PEG. Amyotrophic lateral sclerosis patients generate the most frequent dilemmas concerning timing/efficacy and security of PEG placement.

In neurodegenerative disorders with advanced dementia, there are no data available to support tube feeding versus oral feeding. Patients with advanced dementia are not usually considered appropriate candidates for artificial feeding via PEG tubes.

Legally and ethically, specialized nutritional support should be considered a medical therapy, and advanced directives can help us to resolve conflicts over treatments.
7. References


