

# Dysphagia progression in a MELAS syndrome case: a palliative care perspective

## Evolução da disfagia em um caso de síndrome MELAS: o olhar dos cuidados paliativos

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### ABSTRACT

This article aims to describe a dysphagia progression and a choice of the feeding options in a case of MELAS syndrome, under the perspective of palliative care. It is a case in which a woman at the age of 26 years suffered the first symptoms of the disease and had the swallowing functionality progressively impacted. Speech-Language Therapy follow-up was performed at 6 months with the application of a swallowing safety assessment protocol, Functional Oral Intake Scale (FOIS) and swallowing management, with weekly and monthly outpatient returns. At six months of follow-up, the patient progressed from moderate dysphagia to moderate to severe dysphagia and ranged from levels 5 to 1 of FOIS. The patient maintained oral feeding with consistency restriction, dry swallowing maneuver, and control of volume for liquid intake until the end of the six months of follow-up, when gastrostomy was made. Oral feeding in more than one consistency but with compensations was reduced to exclusive non-oral feeding. We chose to maintain oral feeding until the gastrostomy was placed. Non-suggestion of nasogastric tube was based on the patient's desire and the possibility of oral feeding in at least one food consistency.

**Keywords:** Palliative care; Dysphagia; Brain diseases; Stroke; Tube feeding; Mitochondrial diseases

### RESUMO

Este artigo teve por objetivo descrever a progressão da disfagia e a decisão pela via de alimentação em um caso de síndrome MELAS, sob o olhar dos cuidados paliativos. Trata-se de um caso do sexo feminino, que, por volta dos 26 anos de idade sofreu os primeiros sintomas da doença e teve sua função de deglutição progressivamente impactada. Foi realizado acompanhamento fonoaudiológico durante seis meses, com aplicação do protocolo de Avaliação da Segurança da Deglutição, da *Functional Oral Intake Scale* (FOIS) e gerenciamento da deglutição, com retornos ambulatoriais semanais e mensais. Em seis meses de seguimento, a paciente evoluiu de disfagia moderada a disfagia moderada a grave e variou entre os níveis 5 e 1 da FOIS. Manteve a alimentação por via oral, com restrição de consistências, manobra de deglutições múltiplas e controle de volume para ingestão de líquido, até que, ao final dos seis meses de seguimento, foi realizada gastrostomia. A alimentação por via oral em mais de uma consistência, porém com compensações, foi reduzida a uma alimentação exclusiva por via alternativa, ao longo do acompanhamento fonoaudiológico. Optou-se por manter a via oral de alimentação até a colocação da gastrostomia. A não sugestão de sonda nasogastrica se embasou no respeito à vontade da paciente e na possibilidade de alimentar-se, minimamente, de uma consistência por via oral.

**Palavras-chave:** Cuidados paliativos; Disfagia; Encefalopatias; Acidente vascular encefálico; Alimentação por sonda; Doenças mitocondriais

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## INTRODUCTION

Mitochondria are an organelle present in human cells, with the main function of generating energy. Mutations in mitochondrial DNA, or the genes related to it, lead to mitochondrial dysfunctions, in which there is inability to generate the necessary energy for the organs, especially the nervous system, muscles, kidneys, liver and endocrine system<sup>(1)</sup>.

The syndrome that comprises mitochondrial encephalopathy, lactic acidosis and episodes of stroke is known as MELAS (Mitochondrial myopathy, Encephalopathy, Lactic Acidosis and Stroke), being the most common of the mitochondrial diseases. MELAS syndrome is characterized by episodes of stroke before age 40, encephalopathy and mitochondrial myopathy evidenced by lactic acidosis and morphological impairment of muscle fibers. Most patients present with episodes of stroke, dementia, epilepsy, severe muscle fatigue, exercise intolerance, hemiparesis, visual and auditory alterations, peripheral neuropathy and memory and learning impairment<sup>(1)</sup>. Other symptoms, such as psychiatric manifestations, cerebellar ataxia, involuntary muscle contraction, neuropathy, heart disease, short stature and diabetes mellitus have also been observed in patients with this syndrome<sup>(2)</sup>.

The disease is rare and has a prevalence of 0.18 cases per 100,000 inhabitants per year<sup>(3)</sup>. It presents rapid and aggressive progression<sup>(3)</sup> and the average life expectancy observed is around 16.9 years after the onset of symptoms<sup>(4)</sup>.

During the course of the disease, muscular and neurological alterations can impact the functions of the stomatognathic system, in particular, swallowing.

Speech-Language Therapy (SLT) responsibility lies in the management of swallowing, and in these cases, the importance of an approach based on palliative care principles is emphasized<sup>(5)</sup>, since this function is not only a means of physical or biological maintenance, but also an important form of socialization and pleasure.

According to the World Health Organization (WHO), palliative care encompasses the promotion of the quality of life of patients and family members with life-threatening diseases. Therefore, the promotion of quality of life mainly includes prevention and relief of the individual's suffering<sup>(5)</sup>.

Being a disease of progressive character with an unfavorable diagnosis, it is imperative to undertake a palliative care approach from diagnosis, in order to provide a better quality of life and relief from suffering.

To date, there are few published studies on swallowing, dysphagia, and the decision-making process on the feeding pathway in patients with MELAS syndrome. Therefore, this article aimed to describe the progression of dysphagia and the feeding pathway decision in a case of MELAS syndrome, under the perspective of palliative care.

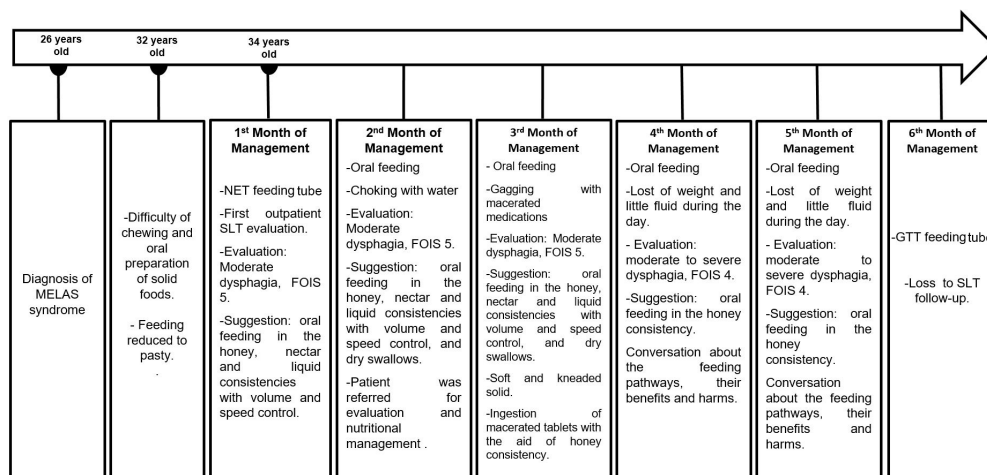
## CASE PRESENTATION

This study respects the Helsinki Resolution and Resolution 466/2012 and was approved by the Research Ethics Committee of the *Universidade Federal de Santa Catarina*, under number 1.316.427. The legal guardian for the patient signed the Terms of Free and Informed Consent.

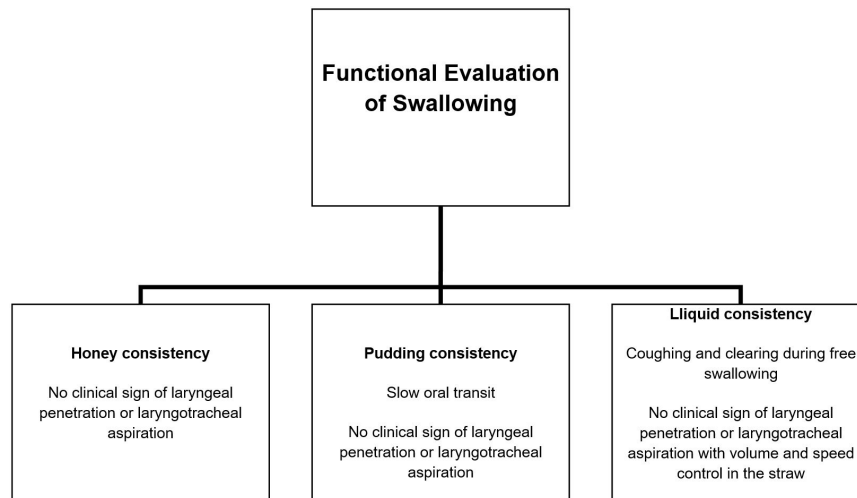
We describe the case of a female patient with MELAS syndrome, 34 years old, Brazilian, single and without children, who at age 26 was diagnosed with progressive mitochondrial encephalopathy, MELAS syndrome.

Eight years after the onset of symptoms, the patient had motor impairment, non-walking and showing generalized weakness in the skeletal muscles, in addition to daytime sleepiness, flaccid dysarthria and difficulty in swallowing. At age 34, the patient was referred by the neurologist to evaluate swallowing and SLT follow-up.

SLT follow-up was initiated based on a swallowing safety assessment protocol<sup>(6)</sup>, the use of compensatory maneuvers to protect the airways and cleanse of stasis after swallowing, changes in diet consistency and control volume by swallowing. The visits were given at the outpatient level with weekly and monthly frequency. The level of oral ingestion at each return was classified by the Functional Oral Intake Scale (FOIS)<sup>(7)</sup> and the severity of dysphagia was based on the Dysphagia Outcome Severity Scale (DOSS)<sup>(8)</sup>. (Figure 1)



**Figure 1.** Chronological representation of the SLT management of swallowing in a case of dysphagia with diagnosis of MELAS syndrome  
**Subtitle:** FOIS - Functional Oral Intake Scale; GTT – gastrostomy; NET – nasogastric tube; SLT – Speech-Language Therapy



**Figure 2.** Clinical signs observed during functional evaluation of swallowing with honey, liquid and pudding consistencies

## Evolution of dysphagia

At age 32, the patient no longer fed on solid consistency. Feeding was spontaneously reduced to pasty, due to the difficulty of chewing and oral preparation of solid foods.

At 34 years of age, the patient had gagged saliva and had been referred for SLT follow-up after the introduction of a nasogastric tube (NET) for alternative feeding during a hospital stay.

## 1<sup>st</sup> Month of management

The patient attended the first outpatient SLT evaluation, moving in a wheelchair, drowsy, communicating in isolated words and responding only to simple verbal commands.

According to the caregiver, the patient had removed the NET on her own accord the previous day and was brought to the medical service to re-establish the alternative feeding pathway.

When asked about the patient's previous history, the caregiver first referred primarily to diarrhea after the introduction of the enteral diet and the need to keep it contained in the bed, so as not to withdraw the NET. The patient then complained of the discomfort caused by the tube. At this time, the patient was at level 1 of FOIS, which represents the exclusive tube feeding.

During the evaluation, cervical control, nasal breathing and efficient lip seal were observed. Absence of salivary sialorrhea or stasis in oral cavity was observed. At rest, the patient presented a contained jaw, non-protruding tongue posture and negative cervical auscultation. The absence of voluntary swallowing of saliva was observed.

The patient's communication was restricted to word vocalizations and small phrases in low intensity, with flaccid dysarthria, characterized by hypernasal, imprecise and pasty speech articulation.

In the structural evaluation, complete dentition and absence of occlusal alteration were observed. The isolated movements of the lips, tongue, cheeks and jaw were minimal and showed imprecision, low amplitude and reduced muscle strength.

The functional evaluation of swallowing was performed with food, in the consistencies\* honey and pudding with soup spoon, liquid in cup (free swallowing) and straw. The results obtained are summarized in Figure 2.

According to the evaluation protocol used, considered as clinical signs of laryngeal penetration or laryngotracheal aspiration were: coughing or clearing before, during or after swallowing; wet voice; positive cervical auscultation; decrease in oxygen saturation; dyspnea; change in skin color, heart or respiratory rate and/or self-reported discomfort.

After the evaluation, it was concluded that the patient had a functional diagnosis of moderate oropharyngeal dysphagia and FOIS level 5. Moderate oropharyngeal dysphagia predicts the risk of aspiration. The patient may eat some consistencies, using specific techniques to minimize aspiration potential and/or facilitate swallowing, requiring supervision<sup>(8)</sup>. At FOIS level 5, oral feeding is recommended in more than one consistency; however, there is a need for special compensation or preparation.

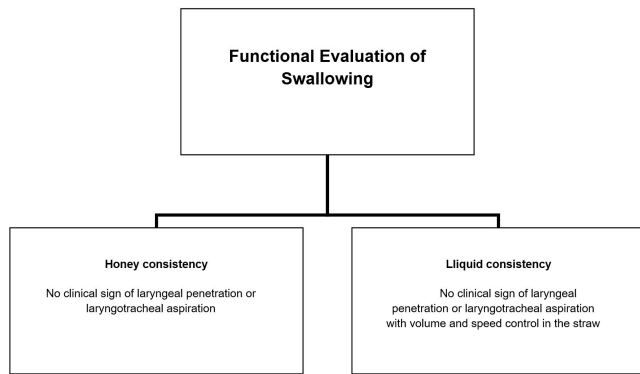
Based on the principles of palliative care<sup>(5)</sup>, the suggestion was to withdraw the NET, as well as to guide oral feeding in the honey, nectar and liquid consistencies. The caregiver was instructed to offer oral feeding, with volume and speed control, namely, offering small sips, with a longer time between them, especially for liquid. It was advised to avoid pudding consistency due to extremely slow oral transit.

The patient was kept under ambulatory monitoring of swallowing. Due to cognitive difficulty, weakness and muscle fatigue, it was decided not to perform exercises or techniques that required greater effort than swallowing itself.

## 2<sup>nd</sup> Month of management

The patient returned, maintaining the clinical profile previously described and oral feeding. The patient's caregiver reported a few incidents of choking with water throughout the week. The consistencies oriented to oral feeding after the

\* According to the American Dietetic Association. National dysphagia diet: standardization for optimal care. Chicago: ADA; 2002.



**Figure 3.** Clinical signs observed during functional assessment of swallowing with honey and liquid consistencies

first evaluation were re-evaluated and the results obtained are presented in Figure 3. After this re-evaluation, the same levels of dysphagia severity and oral ingestion levels were attributed. The same orientations as the previous week were maintained. In addition, it was advised that the caregiver would allow dry swallows before offering a new supply of liquid or food.

The patient was referred for evaluation and nutritional management (since there was no nutritional follow-up initiated at the time of the enteral diet introduction).

### 3<sup>rd</sup> Month of management

After a month, the patient returned, maintaining the initial clinical profile and the caregiver reported gagging only with macerated medications. Regarding diet, there was no reduction in the level of oral intake, or complaints of gagging. The patient expressed the desire to eat rice and beans by mentioning: "I cannot stand to eat everything beaten," referring to eating predominantly in honey consistency.

Based on the principles of palliative care<sup>(5)</sup>, it was advised to offer a soft and kneaded solid, specifically rice and beans, according to the patient's request. Because it was an unsafe consistency, due to the difficulty in the preparatory and oral stages of swallowing, the caregiver was instructed to offer the food in small quantity, only when the patient wished, to be fully alert and seated at 90 degrees. The liquid consistency was maintained under the aforementioned conditions and directed to the ingestion of macerated tablets with the aid of honey consistency.

After this re-evaluation, the levels of dysphagia severity and oral ingestion levels were also maintained.

### 4<sup>th</sup> and 5<sup>th</sup> Months of management

Over the next two months, the patient returned for functional reassessment and swallowing management and was visibly more debilitated, still in wheelchair, drowsy, with longer latency for responses to simple verbal commands and less communicative, as well as remaining with the eyes closed throughout all of the service.

In the 4<sup>th</sup> month of management, the caregiver reported that the patient had lost a lot of weight (reportedly more than 10% of usual body weight), showed no interest in eating, and had little fluid during the day (less than 500 ml, according to information provided by the caregiver).

For functional reevaluation, the same consistencies evaluated previously were used, and slow oral transit and clinical signs suggestive of penetration/aspiration (cough during and after swallowing and wet voice) were observed in all consistencies offered. The consistency with lower risk of aspiration, according to the clinical evaluation, was the honey consistency. In addition, it was observed that the patient performed several incomplete swallows until was able to swallow the bolus. As a course of treatment, it was directed to the caregiver to offer only the honey consistency.

Lastly, moderate to severe oropharyngeal dysphagia and FOIS level 4 were found. In moderate to severe oropharyngeal dysphagia, there is tolerance of only one consistency, with maximum assistance for the use of strategies. There are signs of aspiration of two or more consistencies, requiring multiple clearing requests, absence of reflex cough, weak and ineffective voluntary cough. If the patient's pulmonary status is impaired, it is necessary to suspend oral feeding<sup>(8)</sup>. At FOIS level 4, oral feeding is complete but recommended in only one consistency.

It is noteworthy that pulmonary signs and symptoms were monitored throughout the SLT follow-up and there were no episodes of pneumonia or other pulmonary complications during this period.

It was decided not to submit the patient to videofluoroscopic swallowing study, due to the risk of barium bronchoaspiration, clearly observed during the clinical evaluation. In addition, confirmation of aspiration, or the hypothesis of pathophysiological changes evidenced in the clinical evaluation, would not change the approach adopted.

A conversation was made with the patient and the family about the feeding pathways, their benefits and harms, and then performed a new counter-reference to the neurologist.

### 6<sup>th</sup> Month of management

Continuing to be based on the principles of palliative care<sup>(5)</sup>, the patient remained without reintroduction of NET, with an oral diet oriented towards honey consistency, for two months (4<sup>th</sup> and 5<sup>th</sup> months of management), when at the 6<sup>th</sup> month of follow-up, gastrostomy surgery was performed.

The option for gastrostomy was made by the family, since the patient was considered legally incapable. After gastrostomy, the patient did not return to dysphagia outpatient and contact was lost with the patient's network of care.

## DISCUSSION

The present article proposed the discussion of the evolution of dysphagia and the feeding decision in the case of a 34-year-old female patient with MELAS syndrome, during six months of SLT follow-up.

Swallowing complaints were observed in 48% of the patients with this syndrome, in a cohort of 98 individuals aged between 17 and 85 years. Eighty-eight percent of these patients felt

the food go the “wrong way” at times<sup>(9)</sup>. No publications of longitudinal studies were found that described the evolution of dysphagia in the disease, or that objectively investigated participants’ swallowing.

A case study of an 11-year-old female patient with MELAS syndrome described the loss of functional swallowing (clinically evaluated) since the age of 7 years. With initial impact to the solid consistency and pasty feeding since 7 years of age, it evolved with feeding restricted to liquid and liquid thickened. Four years after the onset of swallowing changes, there was difficulty in opening the mouth, absence of suction and rotational movements of the mandible. As in the present case, the reduction of the volume of the bolus by swallowing was an efficient compensatory maneuver to reduce the clinical signs of laryngotracheal aspiration. There was significant weight loss for the period of one year<sup>(10)</sup>.

The initial swallowing difficulty of the present case was related to the solid consistency. Chewing, oral manipulation, and solid ejection require increased muscle strength, and obviously, increased cellular/muscle energy. Similar to solid, the ejection of the pasty bolus also demands greater muscle strength than the less thick consistencies.

As in the case under discussion, slowed oral transit was also observed in the previously published case study, which may be explained by weakness in the lips, tongue and mandible musculature for the preparation of the bolus, which consequently impairs the conduction of the bolus to the esophagus, raising the risk of pharyngeal residue and laryngotracheal aspiration<sup>(10)</sup>.

Cerebellar ataxia, peripheral neuropathy, muscle weakness and other features of mitochondrial diseases are probably responsible for changes in the pathophysiology of swallowing observed in the case<sup>(9)</sup>.

With the evolution of dysphagia, the liquid consistency also showed a greater risk for aspiration. It is believed that physiologically the liquid consistency requires greater agility and effectiveness in the laryngeal closure, in addition to greater oral motor control, than thickened liquid.

The reduction of the reflexes and movements of the lips, tongue and hyolaryngeal complex justifies the alteration of the sensorimotor skills necessary for swallowing<sup>(10)</sup>, especially with liquid.

Longitudinal studies showed a rapid progression of the symptoms of the disease in cohorts of five<sup>(3)</sup> and ten years<sup>(4)</sup>. The only article found, with a clinical description of the evolution of dysphagia in MELAS syndrome, described a case study. The authors of this study observed a significant impact on the swallowing function over a period of four years and six months of positive responses to SLT follow-up<sup>(10)</sup>.

Unfortunately, the present case delayed the assessment and SLT intervention, and few benefits were observed, except for the adaptation of consistencies and the implementation of compensatory maneuvers during swallowing.

The time elapsed between the beginning of swallowing and of communication difficulty and the beginning of SLT follow-up was one of the factors that compromised the therapeutic process and the performance of palliative care. It is noteworthy that, at present, early follow-up, especially in progressive neurodegenerative diseases, is advocated for the better quality of life, even in the face of a low life expectancy.

There were no reports in the literature regarding the indication or acceptance of an alternative feeding pathway in MELAS syndrome. At present, important and complex bioethical

dilemmas are experienced regarding the decision by procedures and treatments, often considered futile, in cases of patients in exclusive palliative care.

In six months, the change from oral feeding, although with an important restriction of consistency, to an alternative feeding pathway, due to the severity of dysphagia, can constitute a strong change in the individual’s quality of life.

Taking into account, for society as a whole, food has never been seen merely as a means of nutrition, but as an important social, religious, biological and symbolic aspect<sup>(11)</sup>. Thus, the SLT course of treatment established for the case in question were mainly based on the principles of palliative care.

The application of the principles of palliative care in SLT is a topic that has been debated vehemently, especially in the area of dysphagia.

The principles of palliative care include the promotion and relief of pain and suffering; the affirmation of life and the consideration of death as a natural process; the non-acceleration or the postponement of death; the integration of psychological and spiritual aspects in patient care; the provision of supports that enable the patient to live as actively as possible until the moment of his death; the provision of supports that assist the family during the illness, as well as during and after the death of the patient; the offering of multiprofessional support, focusing on the needs of patients and their families, including during mourning; the improvement in the quality of life and positive influence during the course of the disease; the early initiation of palliative care, as well as other measures of prolonged life, including investigations to understand and control stressful clinical situations<sup>(5)</sup>.

In the case under discussion, there was no success in the communication between the teams of the different health services and decisions were not shared. There was a multidisciplinary, but non-interdisciplinary implementation, which compromised compliance with the principles of palliative care, the therapeutic process, maintenance of the oral pathway and control of weight loss. The municipality where the patient resided did not have a home care service, nor have SLT in basic health care. The patient received only medium and high complexity attention at the hospital level. This fact also compromised care and attention to the caregiver and the patient’s family.

The implications of the decision on the feeding pathway, as previously described, prior to initiating SLT follow-up, had the patient using a NET. Both NET and gastrostomy (GTT) allow direct feeding into the gastrointestinal system, without the intersection of this system with the respiratory system. Therefore, it is believed to reduce the risks of bronchoaspiration and pulmonary complications.

The use of NET is considered safe for the patient; however, there are several disadvantages in its use. In addition to being uncomfortable, a factor that has been reported by the patient in question, is that the use of NET causes irritation to the rhinopharynx, the posterior pharyngeal wall, the upper esophageal sphincter and in the long run, may alter local sensitivity. In addition, a NET changes the way glottic closure behaves and does not extinguish bronchospasm, due to the chance of aspiration of saliva, or gastric content. These factors highlight the need to disseminate knowledge and reflections on the subject, so that professionals can clarify all risks to the patient and allow a conscious decision, with better quality of life<sup>(5)</sup>.

Another alternative feeding pathway commonly indicated in cases of dysphagia with unsuccessful therapeutic interventions,

or in cases of use of enteral diet by NET in a period exceeding one month, is a gastrostomy<sup>(12)</sup>.

The advantages of this procedure are easily evidenced when it is thought to protect the patient from health problems such as aspiration pneumonia and malnutrition. However, as in the enteral diet via NET, there are also aspects to be taken into account. The complication rates of its insertion can range from 15% to 70%, with tube leakage, cellulitis, gastroesophageal reflux and diarrhea as some of the possible consequences that may cause extreme discomfort to the patient and a significant worsening of clinical status<sup>(13)</sup>.

For many professionals working in palliative care, the dilemma regarding the use of enteral diet only expands. Physicians, nutritionists and speech-language therapists increasingly question whether the use of such a resource brings benefits to patients in exclusive palliative care. Aggressive nutritional therapies may not be effective, in addition to making treatment more costly and stressful<sup>(14)</sup>.

The difference is also observed that between the mean survival of patients who used gastrostomy and those who did not is not statistically significant in cases of dementia<sup>(15)</sup>. Experts emphasize that the introduction of an alternative feeding pathway should be indicated only when the benefits outweigh the risks and discomforts<sup>(15)</sup>.

Faced with such a respectable and delicate issue, it is important to involve the entire care team, the patient and their family members in order to make decisions about the food pathway.

The need for further studies on swallowing in patients with MELAS syndrome and dilemmas inherent to the feeding pathways is reiterated, since such discussions can make a difference in the care team's conduct, and consequently, in patients' quality of life.

## FINAL COMMENTS

At six months of SLT follow-up, the severity of dysphagia from moderate to severe dysphagia and a significant reduction in the level of oral intake were observed. Oral feeding in more than one consistency, with compensations, has been reduced to exclusive non-oral feeding.

It was decided to maintain oral feeding until the gastrostomy was placed. The non-suggestion of immediate NET was based on respect to the patient's wishes and on the possibility of feeding, at least, a consistency orally.

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