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Case Report
Relato de Caso

Palliative care, amyotrophic lateral sclerosis, and swallowing: a case study

Cuidados paliativos, esclerose lateral amiotrófica e deglutição: estudo de caso

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Keywords

Palliative Care
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ABSTRACT

This study aims to discuss Speech-language pathology (SLP) therapy intervention in dysphagia with a focus on palliative care and quality of life. It is a case study conducted with four participants with amyotrophic lateral sclerosis undergoing SLP therapy outpatient follow-up. The Swallowing Quality of Life Questionnaire (SWAL-QOL) and a structured interview were applied to the participants, who also underwent Videofluoroscopic Swallowing Study (VFSS). Participants were classified according to the Functional Oral Intake Scale (FOIS), the Amyotrophic Lateral Sclerosis Severity Scale, translated and culturally adapted to Brazilian Portuguese (ALSSS), and the Dysphagia Outcome Severity Scale (DOSS). Four patients showed interest in maintaining oral food intake, even if minimal, in the event of tube feeding. Regarding severity of dysphagia, observed in the SVF, the participants presented DOSS classification ranging from functional deglutition to mild-to-moderate dysphagia. The impact on swallowing quality of life was between discrete and severe. Not all participants presented correlation between severity of dysphagia and SWAL-QOL level of impairment, with impact on the quality of life observed even in cases of mild dysphagia severity. Participants reported that they would feel uncomfortable in the event of exclusive tube feeding, and that the oral intake of food, even if minimal only for the pleasure of eating, would have a direct or indirect impact on their quality of life.

Descritores

Cuidados Paliativos
Deglutição
Esclerose Lateral Amiotrófica
Qualidade de Vida
Fonoaudiologia
Disfagia

RESUMO

Tem-se por objetivo discutir aspectos da atuação fonoaudiológica em disfagia, voltada para os cuidados paliativos e a qualidade de vida em deglutição. Trata-se de um estudo de quatro casos com esclerose lateral amiotrófica (ELA) em acompanhamento fonoaudiológico. Foi aplicado o questionário de qualidade de vida em disfagia (SWAL-QOL), realizada entrevista estruturada, classificação da funcionalidade da deglutição pela *Funcional Oral Intake Scale* (FOIS), aplicação da escala de gravidade da ELA (EGELA), realizada videofluoroscopia da deglutição e classificação da severidade da disfagia pela *Dysphagia Outcome Severity Scale* (DOSS). Observou-se que os casos apresentavam tempo de doença entre 12 e 35 meses e possuíam o desejo de manter uma via oral de alimentação, mesmo que mínima, em caso de aceitação da via alternativa de alimentação. Quanto à severidade da disfagia, observada por meio do exame de videofluoroscopia e classificada pela DOSS, apresentavam desde deglutição funcional até disfagia leve a moderada. O impacto na qualidade de vida em deglutição foi mensurado entre discreto e severo. Nem todos apresentavam correspondência entre a severidade da disfagia e a qualidade de vida em deglutição, sendo observado impacto na qualidade de vida, mesmo nos casos com menor grau de disfagia. Os participantes relataram que se sentiriam desconfortáveis em caso de alimentação exclusiva por via alternativa e que a ingestão de alimentos por via oral, mesmo que mínima, apenas pelo prazer da alimentação, refletiria em sua qualidade de vida.

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INTRODUCTION

The World Health Organization defines palliative care as that provided in order to promote the quality of life of individuals suffering from life-threatening diseases by preventing and alleviating physical, psychosocial and spiritual suffering(1). Palliative care employs a way of caring for the process of illness or death without disrespect for the ethics of life or the unnecessary prolongation of suffering(2).

Amyotrophic lateral sclerosis (ALS) is a progressive disease characterized mainly by degeneration of the upper and lower motor neurons. There is an incidence between 2 and 16 new cases per 100,000 individuals(3). Its main signs and symptoms are: progressive weakness, muscular atrophy, fasciculations, muscular cramps, spasticity, dysarthria, dysphagia, dyspnea and emotional liability(4).

As a result of a threat on life, individuals suffering from ALS should be cared for from the outset, that is, from the diagnosis of the disease, by a multidisciplinary team that aims to promote their quality of life. Whenever present, the relief of multidimensional suffering should be the primary focus of the care team.

Life expectancy in ALS is still an object of study and multifactorial, and depends on factors such as clinical presentation, rate of disease progression, early respiratory failure and nutritional status(4). Generally, the individuals affected have a life expectancy around three to five years after the onset of symptoms(3).

Dysphagia is one of the most frequent symptoms in ALS, since the motor nuclei of the IX, X, XI and XII cranial nerve and the corticobulbar tract undergo progressive degeneration, causing a decrease in strength and atrophy of the muscles responsible for swallowing(5).

The change in swallowing directly impacts the function of feeding, which in addition to being a biological need, is a form of social interaction, well-being and personal pleasure. Therefore, the indication of an exclusive alternative feeding pathway should be comprehensively discussed with the team and the patient, with scientific and psychosocial support.

Knowing patients' opinions and their willingness regarding their eating habits may positively influence the care of the speech-language pathologist and the multiprofessional team. Thus, aiming at the quality of life of the patients, the professionals of the team will be able to guide the feeding pathway with less harm and more pleasure, whenever possible.

Based on the foregoing, the objective of this study was to discuss aspects of speech-language pathology (SLP) implementation in dysphagia, focused on palliative care and the quality of life in swallowing of four individuals with ALS.

PRESENTATION OF RESULTS

Procedures performed

This study complied with the Declaration of Helsinki and Resolution 466/2012, and was approved by the Ethics Committee for Human Beings of the *Universidade Federal de Santa Catarina* under number 1,316,427.

This is a study of four cases with medical diagnosis of ALS, in SLP and neurological outpatient follow-up, in a tertiary hospital.

All participants had oral communication, preserved comprehension, exclusive oral food intake and agreed to participate in the research by signing an informed consent form.

Data collection was performed from medical charts, classification of swallowing functionality by the Functional Oral Intake Scale (FOIS)(6), the Amyotrophic Lateral Sclerosis Severity Scale translated version adapted to Brazilian Portuguese (ALSSS)(7), swallowing videofluoroscopy examination, classification of dysphagia severity from the Dysphagia Outcome and Severity Scale (DOSS)(8), dysphagia quality of life questionnaire (SWAL-QOL)(9) and structured interview.

The aim of the interview was to understand the importance of oral feeding for the patient and the desire to maintain an oral intake, even if minimal, in the event of opting for intake by alternative pathway at some point in the progression of ALS. The structured interviews were conducted by the researcher and consisted of open questions and multiple choices, namely: What does food represent for you? (Response options: maintenance of the body, socialization, pleasure or describe another); What is your opinion when you cannot eat some kind of food because you have difficulty swallowing? (Response options: acceptable, unpleasant or unacceptable); Would you like to eat even if mouth feeding is not recommended? (Free response with later guidance was used to describe the flavor, temperature, frequency of food supply and whether they would like something specific).

The SWAL-QOL, translated version adapted to Brazilian Portuguese(9), was used. This instrument aims to evaluate the impact of swallowing changes on the individual's quality of life. It consists of forty-four questions with responses in Likert-type scale, separated into eleven domains: *Swallowing as a Burden*, *Desire to Eat*, *Feeding Duration*, *Frequency of Symptoms*, *Food Selection*, *Communication*, *Fear of Eating*, *Mental Health*, *Social*, *Sleep* and *Fatigue*. Scoring ranges from 0 to 100 (0=worse, 100=better) in the quality of life in swallowing. For the present study, the score was calculated by domain and overall. For the overall score, the scores from 0 to 49 were considered as severe impact, from 50 to 70 as moderate impact and from 71 to 100 as discrete or non-impact.

The translated version of the Amyotrophic Lateral Sclerosis Severity Scale adapted to Brazilian Portuguese (ALSSS)(7) was also used. It is a severity scale of ALS, composed of four dimensions: lower extremity, upper extremity, speech and swallowing. Each can be measured from 1 to 10, one being the worst feature and 10 being the best. Ultimately, the sum of the dimension scores is calculated for the total score calculation. The closer to 40 points, the better the overall functionality. Considered for classification on the scale was neurological, SLP and videofluoroscopic evaluation data, along with complaints presented by the patient.

In order to classify the functionality of participants' swallowing, the FOIS(6) was applied, which classifies the level of oral intake into seven levels. For the present study, it was adapted as follows: Level 1 - nothing by oral pathway; Level 2 - dependent on feeding by alternative pathway with minimal offer of food by mouth; Level 3 - dependent on alternative feeding pathway with constant oral intake of food or liquids; Level 4 - total oral pathway diet in only one or two consistencies (e.g., nectar and honey, honey and pudding); Level 5 - total oral pathway diet with multiple consistencies, but requiring special preparation or compensations; Level 6 - total oral pathway diet with multiple consistencies without special preparation, but with specific food limitations (e.g., fiber, grains and vegetables) and modification of speed and volume if necessary; Level 7 - total oral pathway diet without restrictions.

Swallowing videofluoroscopy examination was performed by a radiologist, a specialized technician and a speech-language pathologist. In the examination, three offers of consistencies were made: liquid (in free swallow), honey and pudding (all in tablespoon) and solid (on demand). The consistencies were obtained as follows: 30 ml of barium (BaSO4) to 20 ml of water for liquid; 20 ml of barium to 15 ml of water and a tablespoon of food thickener to pudding; a biscuit of water and salt soaked in barium for the solid consistency. For the honey consistency, the barium itself was used.

Food was offered to the participants in the following sequence: liquid, honey, pudding and solid. The participants were placed in a sitting position (lateral and anteroposterior) in a special chair, adapting posture when needed. The examination was performed with a remote-controlled sero-apparatus and recorded for later analysis.

For classification according to the DOSS, the results of the exam and the clinical history of each case were qualitatively analyzed by consensus of two SLP therapists specialized in dysphagia.

CASE 1

Case 1, male, 50 years of age. The patient started with symptoms of imbalance followed by falls 18 months prior. Later, the patient noticed a sensation of "food stopped in the throat". No other comorbidities were reported (Table 1).

Table 1. Descriptive analysis of clinical aspects and swallowing quality of life of 4 cases with amyotrophic lateral sclerosis

Variables	CASE 1	CASE 2	CASE 3	CASE 4
Disease duration (months)	18	24	12	35
First symptoms	appendicular (LLs)	appendicular (LLs)	bulbar (speech and swallowing)	appendicular (LLs)
Motor limitations	Changes observed in gait	Changes observed in gait	Weakness in ULs	Walking with mechanical apparatus
Respiratory aspects	Dyspnea (in nocturnal use of BIPAP)	Nothing noteworthy	Nothing noteworthy	Nothing noteworthy
Psychoemotional aspects	Medical diagnosis of depression	Medical diagnosis of depression	Easy crying	Irritation and emotional instability
ALSSS Scale	34	34	30	33
Lower limbs	7	7	10	6
Upper limbs	10	10	9	9
Speech	9	9	5	9
Swallowing	8	8	6	9
Other comorbidities	Denied	SAH	Denied	SAH
Family history	Denied	Denied	Yes	Denied
SWAL-QOL domains				
Swallowing as a burden	12	62	50	100
Eating desire	66	41	50	91
Eating duration	0	0	0	100
Frequency of symptoms	51	66	66	91
Food selection	87	50	62	62
Communication	12	25	25	87
Fear of eating	69	81	6	94
Mental health	25	100	50	90
Social	50	40	75	100
Sleep	37	50	100	75
Fatigue	100	83	75	100
Overall SWAL-QOL score	Severe	Moderate	Moderate	Mild
FOIS	5	7	5	6
Severity of dysphagia (DOSS)	Mild	Mild	Mild to moderate	Functional swallowing
Representation of feeding	Socialization	maintenance of body	maintenance of body	socialization
No oral intake	unpleasant	unpleasant	unacceptable	unpleasant
Oral intake taste	Salty	Salty	Salty	Salty
Oral intake temperature	Hot	Room temperature	Hot	Hot
Oral intake frequency	4x/day or +	4x/day or +	2x/day	3x/day

Caption: ULs = Upper limbs; LLs = Lower limbs; BIPAP = *Bi-level Positive Pressure Airway*; SAH = Systemic arterial hypertension; ALSSS = *Amyotrophic Lateral Sclerosis Severity Scale*; SWAL-QOL = *Swallowing Quality of Life Questionnaire;* FOIS = *Functional Oral Intake Scale*; DOSS = *Dysphagia Outcome Severity Scale*

At that time, the patient reported the use of drugs that could cause dry mouth (as a side effect), as well as antidepressants and complained of worsening dyspnea and dysphagia.

In swallowing videofluoroscopy, mild oropharyngeal dysphagia(8) was observed, with a significant presence of pharyngeal residues, but with absence of laryngeal penetration or laryngotracheal aspiration. The patient was at level 5 of the FOIS.

In the interview, the patient said that food represented a form of socialization and that the absence of oral feeding would be unpleasant, but not unacceptable. The patient reported that he would like to put some food in his mouth just for pleasure, in case of exclusive feeding by alternative pathway. The patient showed a preference for salty, hot food and was offered more than four times a day.

According to the overall SWAL-QOL score, the patient presented severe impairment of the quality of life in swallowing. The questionnaire score by domains is shown in Table 1.

CASE 2

Case 2, male, 61 years of age. The patient reported weakness in the lower limbs 24 months prior, and in the last months, noticed difficulty speaking. The patient also reported hypertension and depression (Table 1).

In swallowing videofluoroscopy, mild oropharyngeal dysphagia(8) was observed, with presence of residues in oral cavity and vallecula after swallowing, but with no laryngeal penetration or laryngotracheal aspiration. The patient was at level 7 of the FOIS.

In the interview, the patient said that food represented a way of maintaining the body and that the absence of oral feeding would be unpleasant, but not unacceptable. The patient reported that he would like to put some food in his mouth just for pleasure, in case of exclusive feeding by alternative pathway. The patient showed a preference for salty food at room temperature and was offered more than four times a day.

According to the overall SWAL-QOL score, the patient presented moderate impairment of the quality of life in swallowing. The questionnaire score by domains is shown in Table 1.

CASE 3

Case 3, female, 29 years of age. The patient began with difficulty speaking and swallowing 12 months prior, with significant limitation of tongue movements, and more recently, noted weakness in the upper limbs. At that time, the patient reported improvement of dysphagia with the use of therapeutic bandaging applied by the SLP therapist. The patient also presented a history of the disease in the family (Table 1).

In swallowing videofluoroscopy, mild to moderate oropharyngeal dysphagia was observed(8). The patient presented absence of lip seal and extra-oral escape, in addition to slowness and inefficiency of tongue movements and chewing. There was presence of residues in oral cavity, vallecula and pyriform recess after swallowing for all consistencies offered. Reduced laryngeal elevation and an episode of laryngeal penetration during swallowing of liquid could also be observed. The patient was at level 5 of the FOIS.

In the interview, the patient reported that food represented a way of maintaining the body and that the absence of oral feeding would be unacceptable. The patient said she would like to put some food in her mouth just for pleasure, in case of exclusive feeding by alternative pathway. The patient showed preference for hot, salty food offered twice a day, in addition to not revealing an option for any specific food.

According to the overall SWAL-QOL score, the patient presented moderate quality of life in swallowing (Table 1).

CASE 4

Case 4, male, 52 years of age. The patient started with weakness in the lower right limb for 35 months and, six months later, noticed difficulty speaking and reported gagging and occasional falls. The patient reported hypertension as comorbidity (Table 1).

In the period proximate to participation in the study, the patient reported an increase in the number of falls, irritation and emotional instability.

After swallowing videofluoroscopy, functional swallowing was observed(8) with a discrete presence of residues in the oral cavity, vallecula and pyriform recess after swallowing, but without laryngeal penetration or laryngotracheal aspiration. The patient was at level 6 of the FOIS.

In the interview, the patient said that food represented a form of socialization and that the absence of oral feeding would be unpleasant, but not unacceptable. The patient reported that he would like to put some food in his mouth just for pleasure, in case of exclusive feeding by alternative pathway. The patient showed preference for hot, salty food, offered three times a day.

According to the overall SWAL-QOL score, there was a slight impairment of the quality of life in swallowing (Table 1).

Although it was discussed with the first three cases, the future possibility of insertion of an alternative feeding pathway, was not indicated for any of the individuals.

DISCUSSION

ALS is a disease of rapid progression and throughout the process of degeneration, there is an unavoidable impairment of oral feeding. When discussing palliative care and swallowing, it is understood that it is necessary to take into account ways to adapt swallowing, with the objective of maintaining the pleasure of oral feeding, with the maximum possible safety, but mainly, maintaining the quality of life of the individual.

The quality of life in swallowing is conceived as complex and individualized. Only when we consider the individual's perception of their own life and about themselves will there be a chance to establish actions with an effective positive impact.

Although alternative food intake has not yet been indicated for the cases under discussion, almost invariably this is recommended in patients with ALS. The indication of an alternative intake pathway (usually gastrostomy) in ALS is initially due to malnutrition, dehydration and weight loss. Weight loss is a common occurrence not only of dysphagia, but also of increased energy expenditure caused by muscle fasciculations, hypermetabolism, and increased respiratory effort. Additionally, depression is often associated and leads to decreased appetite and the oral intake of food(10).

According to the Progas Study Group(11), in the cases of ALS, the sooner a gastrostomy (as an alternative or supplementary intake pathway) is performed, the greater the chances of recovery of weight and increased survival of the individual. However, according to the same study, the effect of gastrostomy on patients' quality of life may not be significant. Quality of life may not be necessarily or absolutely associated with the absence of hunger or with the total supply of the caloric and hydration needs of individuals with ALS.

In the cases under discussion, there was no complete correspondence between swallowing functionality and the swallowing quality of life index, and this observation differed from studies on swallowing quality of life, with participants suffering from dysphagia due to non-degenerative diseases(12,13).

It was observed that Case 1, even with mild dysphagia and FOIS 5, presented the worst index of quality of life in swallowing of the analyzed cases. Case 3, which showed a higher degree of dysphagia and an equal level of intake, observed a moderate impact on the quality of life. With equal impact on quality of life, Case 2 did not present the same degree of impairment in swallowing functionality.

Case 1 also obtained a considerably reduced score in SWAL-QOL compared to other participants in the following domains: *Swallowing as a Burden*, *Communication*, *Mental Health*, *Social* and *Sleep*. This case had a diagnosis of depression and was the only one with significant respiratory impairment. Although with ALSSS at a level close to normal, the impairment observed in the SWAL-QOL and respiratory distress may have negatively influenced the quality of life in swallowing.

In the interview, this same participant reported considering feeding as a form of socialization, having presented reduced scores in the social domain of SWAL-QOL. In this case, the influence of communication difficulty on the quality of life in swallowing was also observed, although other people did not notice changes in their speech.

Case 4 was the participant with swallowing quality of life and swallowing functionality, empirically, more correlated. The participant presented functional swallowing, FOIS 6, and a discrete impact on quality of life. The most affected SWAL-QOL domain in this case was *Food Selection,* in which knowing what to eat or not can sometimes be a problem for the participant.

Case 2, even considering feeding only as a form of body maintenance, would like to receive some kind of oral feeding only for pleasure, more than four times a day. Nevertheless, the participant presented the lowest score in the domain desire to feed in the SWAL-QOL in relation to the other cases. Since this case presented the medical diagnosis of depression, it is important to consider the impact of this comorbidity on appetite and the desire to eat(10).

Only Case 3 considered it unacceptable not to be able to eat orally, who defined the diet only as a way of maintaining the body. This case was the only one that demonstrated bulbar alterations as early symptoms and presented airway permeation during the instrumental evaluation of swallowing. It is questioned whether this participant's difficulty in swallowing and frequent gagging has led her to the meaning of feeding herself only to maintain the body.

In this case, the domain *Fear of Feeding* from the SWAL-QOL presented an extremely low score, indicating the impact of this aspect on quality of life. Still, the participant said that losing the ability to eat orally would be unacceptable and would compromise her autonomy even more.

Even though there was no change in gait and few appendicular symptoms, the ALSSS classification was the lowest, which expresses the magnitude of bulbar impairment in daily functionality.

This case was the only one with a diagnosis of familial ALS. Traditionally, ALS can be classified as sporadic or familial. Family cases account for 10% of the disease incidence rate and are more associated with the early onset of symptoms(14). In cases with a family history, the suffering may be even greater, given the fact of having accompanied a loved one during the progression of the disease, observing the concrete and total loss of functionality. Failure to feed orally caused by severe dysphagia ends up being a sign of the inevitable progress of the disease.

The conception and the representation of feeding were one of the factors that also influenced the quality of life in swallowing. Food represents more than just the nutritional aspect, especially for individuals in palliative care. It is a motivation for living with illness, showing affection, compassion, and even, accepting death(15).

Patients can signify food as a source of energy to maintain the body and stay in the fight against the disease. But they can also identify this as a source of frustration and anxiety or the care and concern of the team's professionals for them. Therefore, to provide quality of life in palliative care, it is necessary to address the individual's meaning of feeding(15).

In view of the fact that all the participants stated that they wished to maintain oral feeding, albeit at minimum for pleasure (i.e., in the event of the inability to feed by mouth due to severe dysphagia), it is important to take into account the patients' desire and the impact that suspension of oral intake can have on their quality of life.

All participants reported preferring that the food offered was at room temperature or warm, which is believed to be related to the season (winter) in which the study was conducted. Although there is evidence that ice and sour foods are the most suitable for triggering swallowing in neurological cases, for patients in palliative care, it is suggested that the actions of SLP therapy also reflect the desire of the individual, who will often have a small oral intake of food.

Often the consequences of curative processes in patients with advanced and/or terminal diseases is the prolongation of suffering through interventions and treatments that do not generate substantial benefits for patients(2).

It is essential to take into account that the patient does not only present a physical/biological problem, and therefore must be attended to with a holistic view, aiming above all quality of life.

There are few published studies on palliative care and swallowing in patients with neurodegenerative diseases, especially on the SLP therapist's performance with these patients. Therefore, the importance of observational and experimental studies on SLP in palliative care and neurodegenerative diseases is emphasized.

FINAL CONSIDERATIONS

The cases studied considered swallowing as a form of socialization or maintenance of the body. They ranged from functional swallowing to mild to moderate dysphagia, with an impact on the quality of life from mild to moderate swallowing. There was no correlation between dysphagia severity and swallowing quality of life in all cases, with a higher impact on quality of life observed, even in cases with a lower degree of dysphagia. It was observed that other factors were intrinsically associated with the quality of life in swallowing, with not only its functionality.

It is important to know the patients' desire for their food and the meaning they attribute to it, so that the SLP therapy can actually contribute to the quality of life of the individual. It is believed that when considering the multidimensional manifestations of ALS, the quest for quality of life from the principles of palliative care and pleasurable feeding may reflect positively throughout the progress of the disease.

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Authors contributions

KFL contributed with study design, analysis and interpretation of data, writing and review of the manuscript; ICS contributed with the collection and analysis of the data and writing of the manuscript.