KAREN FONTES LUCHESI

DYSPHAGIA IN AMYOTROPHIC LATERAL SCLEROSIS AND PARKINSON'S DISEASE

A DISFAGIA NA ESCLEROSE LATERAL AMIOTRÓFICA E NA DOENÇA DE PARKINSON

CAMPINAS

2013

UNIVERSIDADE ESTADUAL DE CAMPINAS

FACULDADE DE CIÊNCIAS MÉDICAS

KAREN FONTES LUCHESI

DYSPHAGIA IN AMYOTROPHIC LATERAL SCLEROSIS AND PARKINSON'S DISEASE

Orientador: Prof. Dr.Satoshi Kitamura

A DISFAGIA NA ESCLEROSE LATERAL AMIOTRÓFICA E NA DOENÇA DE PARKINSON

Tese de doutorado apresentada à Pós-Graduação em Saúde Coletiva da Faculdade de Ciências Médicas da Universidade Estadual de Campinas, para obtenção do título de doutora em Saúde Coletiva, área de concentração em Epidemiologia.

Doctorate thesis presented to the Collective Health Postgraduation Programme of the School of Medical Sciences of the University of Campinas to obtain the Ph.D grade in Collective Health, Epidemiology.

ESTE EXEMPLAR CORRESPONDE À VERSÃO FINAL DA TESE DEFENDIDA PELA ALUNA KAREN FONTES LUCHESI E ORIENTADO PELO PROF. DR. SATOSHI KITAMURA.

Assinatura do Orientador

CAMPINAS

2013

FICHA CATALOGRÁFICA ELABORADA POR
MARISTELLA SOARES DOS SANTOS – CRB8/8402
BIBLIOTECA DA FACULDADE DE CIÊNCIAS MÉDICAS
UNICAMP

L963d

Luchesi, Karen Fontes, 1984-
A disfagia na esclerose lateral amiotrófica e na doença de Parkinson / Karen Fontes Luchesi . -- Campinas, SP : [s.n.], 2013.

Orientador : Satoshi Kitamura.
Tese (Doutorado) - Universidade Estadual de Campinas, Faculdade de Ciências Médicas.

1. Doenças neurodegenerativas. 2. Deglutição. 3. Fonoterapia. I. Kitamura, Satoshi, 1945-. II. Universidade Estadual de Campinas. Faculdade de Ciências Médicas. III. Título.

Informações para Biblioteca Digital

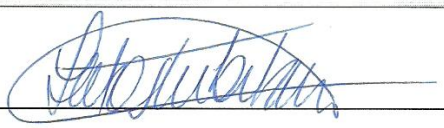
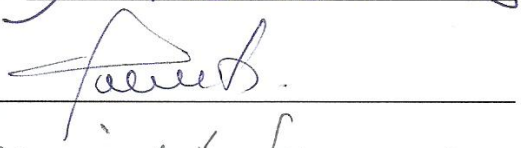
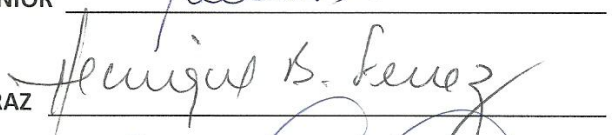
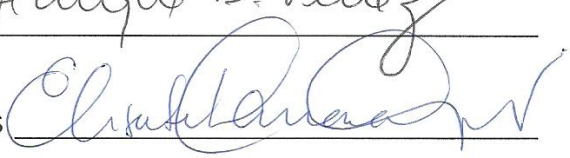
Título em inglês: Dyphagia in Amyotrophic Lateral Sclerosis and in Parkinson's Disease.
Palavras-chave em inglês:
Neurodegenerative diseases
Deglutition
Speech Therapy
Área de concentração: Epidemiologia
Titulação: Doutora em Saúde Coletiva
Banca examinadora:
Satoshi Kitamura [Orientador]
Maria Rita Donalisio Cordeiro
Marcondes França Junior
Henrique Ballalai Ferraz
Elisabete Carrara de Angelis
Data da defesa: 06-02-2013
Programa de Pós-Graduação: Saúde Coletiva

BANCA EXAMINADORA DA DEFESA DE DOUTORADO

KAREN FONTES LUCHESI

Orientador (a) PROF(A). DR(A). SATOSHI KITAMURA

MEMBROS:

1. **PROF(A). DR(A). SATOSHI KITAMURA** 
2. **PROF(A). DR(A). MARIA RITA DONALISIO CORDEIRO** 
3. **PROF(A). DR(A). MARCONDES FRANÇA JUNIOR** 
4. **PROF(A).DR(A). HENRIQUE BALLALAI FERRAZ** 
5. **PROF(A).DR(A). ELISABETE CARRARA DE ANGELIS** 

Programa de Pós-Graduação em Saúde Coletiva da Faculdade de Ciências Médicas da Universidade Estadual de Campinas

Data: 06 de fevereiro de 2013

AGRADECIMENTOS

Agradeço a Deus por esta prazerosa e importante conquista.

Ao Prof. Dr. Satoshi Kitamura pelos anos de orientação, apoio, encorajamento e compreensão.

À Profa. Dra. Lucia Figueiredo Mourão pelos anos de orientação, incentivo, compreensão, encorajamento, amizade e exemplo.

À Jeanine Ma pela laboriosa e valiosa revisão da língua inglesa.

Aos pacientes que possibilitaram a realização esta pesquisa e contribuíram para a construção do conhecimento.

Ao meu esposo Ezequiel pelos quatro anos de amor, compreensão, apoio e paciência.

Aos meus pais, que desde pequena me ensinaram a ter sede de conhecimento, agradeço pelo amor, incentivo e dedicação.

Ensina-nos a contar os nossos dias,

para que alcancemos coração sábio.

(Bíblia Sagrada, Salmos 90:12)

RESUMO

A Doença de Parkinson (DP) é uma das doenças neurodegenerativas mundialmente mais prevalentes. Dentre as doenças do neurônio motor, a Esclerose Lateral Amiotrófica (ELA) é a mais frequente. A qualidade de vida e o prolongamento da expectativa de vida dos sujeitos com doenças neurodegenerativas, como ELA e DP, são foco da intervenção fonoaudiológica, visto que uma das maiores causas de morte são as pneumonias aspirativas. Esta pesquisa teve por objetivo analisar e descrever aspectos relacionados à disfagia e à sua progressão em sujeitos diagnosticados com ELA e DP. Ao todo, participaram 49 sujeitos com ELA e 24 sujeitos com DP. Todos foram avaliados e acompanhados no ambulatório de Otorrinolaringologia/Disfagia do Hospital de Clínicas da Universidade Estadual de Campinas. Foram incluídos no estudo, apenas os sujeitos que estavam em acompanhamento periódico no ambulatório de neurologia do referido hospital e em tratamento medicamentoso. Foram excluídos os sujeitos sem queixa de deglutição ou que apresentassem outras doenças que pudessem causar alteração na deglutição. Todos foram submetidos à entrevista estruturada, videoendoscopia da deglutição, avaliação clínica da deglutição e intervenção fonoaudiológica, além de terem a funcionalidade de ingestão oral classificada pela *Functional Oral Intake Scale*. Foi realizada uma análise descritiva dos dados com apresentação de frequência das variáveis categóricas e medidas de tendência central e dispersão das variáveis numéricas. Na análise exploratória foram utilizados: Regressão de Cox, teste Exato de Fisher, teste de Kruskal-Wallis, Qui-quadrado, teste de Mann-Whitney e análise de sobrevivência de Kaplan-Meier. As análises foram realizadas por meio do *software* SPSS versão 13.0 para Windows, tendo sido adotado o nível de significância para os testes estatísticos de 0,05. Na ELA, foi identificado como fator associado à disfagia moderada ou grave, a odinofagia (p=0,01). Foram identificados como fatores que

influenciaram na progressão da disfagia na ELA a idade de início da doença (p=0,02) e o início bulbar (p=0,04). A idade de início avançada (p=0,03) e o menor tempo de doença até a primeira avaliação (p=0,004) foram identificados como fatores que levaram à necessidade de indicação de uma via alternativa de alimentação em menor tempo na ELA. Não foram identificados fatores que influenciassem a progressão da disfagia na DP. Observou-se melhora e estabilização da função de deglutição na maioria dos sujeitos com DP estudados. Conclui-se que a idade de início e o início bulbar da ELA são fatores associados à piora rápida da disfagia. Não houve fatores associados à progressão da disfagia na PD e a funcionalidade na deglutição destes pacientes foi caracterizada por melhora e manutenção.

Palavras-chave: Doenças neurodegenerativas, deglutição, fonoterapia.

ABSTRACT

Parkinson's disease (PD) is one of the most prevalent neurodegenerative diseases worldwide. Among the motor neuron diseases, the Amyotrophic Lateral Sclerosis (ALS) is the most common. The quality of life and longer life expectancy for these individuals with neurodegenerative diseases are the purpose of speech-language therapy, as the leading cause of death are aspirative pneumonias. The objective of this study was to analyze and describe aspects related to dysphagia and its progression in patients diagnosed with ALS and PD. Altogether, 49 patients with ALS and 24 patients with PD participated in the study. All patients were evaluated and followed by at the Otolaryngology/Dysphagia services of the Clinical Hospital of the University of Campinas. The study included only patients who have been regularly monitored at the neurology service and undergoing drug treatment. Patients who had other conditions that could cause changes in swallowing or with no complaints concerning swallowing were excluded. All patients underwent a structured interview, Fiberoptic Endoscopic Evaluation of Swallowing, clinical evaluation of swallowing and swallowing management. Furthermore, they had the swallowing functionality classified by the Functional Oral Intake Scale. We performed a descriptive analysis with presenting the frequency of categorical variables, central measures tendency and dispersion of numerical variables. In exploratory data analysis were applied Cox Regression, Kruskal-Wallis, Chi-square, Mann-Whitney and survival analysis of Kaplan-Meier. The analyses were performed using SPSS version 13.0 for Windows and the significance level for statistical tests was 5%. Odynophagia was identified as an associated factor with moderate or severe dysphagia in ALS. The onset age of ALS ($p = 0.02$) and the

bulbar onset ALS ($p = 0.04$) were identified as factors that influence the progression of dysphagia in ALS. Advanced onset age ($p = 0.03$) and shorter disease duration ($p = 0.004$) were identified as factors that lead to sooner need for non-oral feeding. We did not identify any associated factors with the progression of dysphagia in PD. We noticed an improvement and stabilization of the swallowing function in most patients with PD. We conclude that the onset age and bulbar onset of ALS are factors associated with rapid worsened dysphagia.

Key-words: Neurodegenerative Diseases, Deglutition, Speech Therapy

LISTA DE ABREVIATURAS

ALS	*Amyotrophic Lateral Sclerosis*
ELA	Esclerose Lateral Amiotrófica
PD	*Parkinson's Disease*
DP	Doença de Parkinson
FEES	*Fiberoptic Endoscopic Evaluation of Swallowing*
VED	Videoendoscopia da Deglutição
FOIS	*Functional Oral Intake Skale*

LISTA DE TABELAS

CAPÍTULO 1

CAPÍTULO 2

CAPÍTULO 3

LISTA DE FIGURAS

CAPÍTULO 2

CAPÍTULO 3

SUMÁRIO

INTRODUÇÃO E OBJETIVOS

INTRODUÇÃO

As doenças neurodegenerativas apresentam em seu curso, alterações no processo da deglutição. A disfagia é um dos sintomas de importante risco para a vida dos pacientes acometidos, especialmente, na presença de tosses e engasgos durante ou após a alimentação, sensação de alimento parado, sialorréia, perda de peso e pneumonia aspirativa.[1]

A Doença de Parkinson (DP) é uma das doenças neurodegenerativas mundialmente mais comum, com prevalência de 1,1 a 1,7 por 1000 na população geral, aumentando com a idade.[2] Apresenta incidência anual de 1 a 20 por 1000 pessoas.[3] Por outro lado, entre as doenças do neurônio motor, a Esclerose Lateral Amiotrófica (ELA) é a mais frequente, com uma incidência anual mundial de 1 a 3 por 100.000 pessoas[4] e prevalência de 2,7 a 7,4 em 100.000 pessoas por ano.[5] Por isso, optou-se por estudar a disfagia nestas doenças.

De acordo com a literatura, os pacientes com ELA tem uma expectativa de vida de aproximadamente 3 a 5 anos após o início dos sintomas.[6] Tem como uma das principais causas de morte as pneumonias aspirativas.[6,7] A ELA é caracterizada pela degeneração dos neurônios motores, tronco cerebral e medula.

A disfagia na ELA tem caráter progressivo e origina-se na fraqueza e espasticidade dos músculos inervados pelos nervos trigêmeo, facial, hipoglosso, glossofaríngeo e vago.[8] Na ELA a espasticidade, a atrofia e o tônus muscular reduzido podem causar dor e diminuir ainda mais a coordenação e a função dos músculos afetados.[9]

A indicação de via alternativa de alimentação na ELA é bastante comum nos estágios mais avançados da doença, nos quais a disfagia grave, a fadiga muscular e/ou a insuficiência respiratória dificultam, ou impossibilitam, que todo o aporte alimentar seja ingerido por via oral. A inserção de vias alternativas de alimentação em tempo adequado pode auxiliar na prevenção de pneumonias aspirativas e desnutrição, de modo a propiciar aumento da sobrevida nos pacientes acometidos.

A DP, por sua vez, é caracterizada pela afecção dos gânglios da base, causando diminuição dos movimentos voluntários, tremor de repouso, rigidez, bradicinesia e inabilidade postural.[10,11]

A disfagia é um sintoma bastante comum em pacientes com DP, de acordo com a literatura afeta mais de 80% desses pacientes e é progressiva assim como o curso natural da doença.[12] As alterações de deglutição na DP estão frequentemente associadas às fases oral e faríngea devido à dificuldade de manipulação do bolo alimentar e atraso no disparo do reflexo de deglutição.[13]

Na DP há uma expectativa de vida de aproximadamente 14 anos após o aparecimento dos primeiros sintomas da doença[14] e as complicações da disfagia, como a desnutrição e a pneumonia aspirativa, estão comumente associadas ao aumento da mortalidade desses pacientes.[15,16]

O acompanhamento fonoaudiológico faz-se importante em doenças neurodegenerativas como ELA e DP, pois auxilia na adaptação das funções de alimentação e comunicação. Em um passado não muito distante, profissionais, pacientes e familiares acreditavam que por haver uma piora inevitável nessas funções devido ao caráter progressivo das doenças, o acompanhamento fonoaudiológico era dispensável.

Existe uma escassez de publicações que evidenciam a evolução da disfagia nos pacientes com ELA e DP. Há estudos sobre a caracterização e o tratamento da disfagia na ELA e na DP[6, 12, 14, 16, 17, 18, 19, 20, 21, 22, 23, 24, 25]. No entanto, não foram encontrados na literatura, nacional ou internacional, muitos estudos sobre a progressão da disfagia na DP e na ELA. Até o momento, foi identificada apenas uma série de casos acompanhada no Japão em que os autores descrevem alguns aspectos da evolução da disfagia na ELA.[26] Não foram encontrados estudos sobre o acompanhamento da progressão da disfagia na DP.

O presente estudo tem por objetivo descrever a evolução da disfagia e os fatores que podem influenciar a sua evolução, sob cuidado ambulatorial, ao longo do tempo.

Trata-se de um estudo longitudinal realizado durante um período de cinco anos (2006-2011) com pacientes do Ambulatório de Otorrinolaringologia/Disfagia do Hospital de Clínicas da Universidade Estadual de Campinas (Unicamp). Neste ambulatório, a coleta de dados se dá por meio padronizado, as avaliações seguem o protocolo já publicado por Sordi et al.[27] (ANEXO I) e os retornos são registrados segundo o roteiro anexo (ANEXO II).

Esta tese é composta por três artigos. O primeiro discute os fatores associados à severidade da disfagia na ELA. O segundo traz uma análise de sobrevivência para discutir os fatores que podem influenciar no tempo decorrido até a indicação de uma via alternativa ou à piora da disfagia na ELA. Já o terceiro apresenta uma análise descritiva sobre a progressão da disfagia e a intervenção fonoaudiológica em cinco anos de acompanhamento de pacientes com DP.

OBJETIVOS DO ESTUDO

OBJETIVO GERAL

Descrever e analisar os aspectos clínicos e evolutivos relacionados à disfagia em sujeitos diagnosticados com Esclerose Lateral Amiotrófica (ELA) e Doença de Parkinson (DP).

OBJETIVOS ESPECÍFICOS

1. Identificar fatores associados à gravidade da disfagia em sujeitos com ELA;
2. Identificar fatores associados ao intervalo de tempo entre o início dos sintomas e a progressão da disfagia e a necessidade da inserção de via alternativa de alimentação em sujeitos com ELA;
3. Realizar análise descritiva sobre a progressão da disfagia e a intervenção fonoaudiológica ambulatorial em sujeitos com DP.

CAPÍTULO 1

ASSOCIATED FACTORS WITH DYSPHAGIA SEVERITY: ODYNOPHAGIA IN AMYOTROPHIC LATERAL SCLEROSIS

Fatores associados à gravidade da disfagia: odinofagia na Esclerose Lateral Amiotrófica

Autores: Luchesi KF, Kitamura S, Mourão LF.

(Submetido)

Abstract

Purpose: This article aims to identify associated factors with dysphagia severity in Amyotrophic Lateral Sclerosis (ALS).

Method: It is a cross-sectional study of 49 patients with ALS. All patients answered a verbal questionnaire about swallowing complaints and underwent Fiberoptic Endoscopy Evaluation of Swallowing. The patients were divided into groups according to dysphagia severity. At the clinical evaluation the patients' current age at the assessment, age at onset of ALS symptoms, the complaints of oral and pharyngeal phase of swallowing were identified. To identify associated factors with dysphagia severity in ALS, Cox Regression was used. The analysis were performed using SPSS version 13.0 for Windows and statistical tests considered p-values less than or equal to 0.05.

Results: The higher number of swallowing complaints of oral and pharyngeal phase (p-value: 0.03 and 0.009), the reference of difficulty to eject the bolus (p-value: 0.03) and odynophagia (painful swallowing) (p-value: 0.01) were aassociated with moderate or severe dysphagia. After differential analysis, only odynophagia remained as an associated factor of moderate or severe dysphagia (risk: 11.96; p-value: 0.01).

Conclusion: The odynophagia was associated with moderate and severe dysphagia, in ALS suggests a high risk of pulmonary and nutritional complications.

Key words: Amyotrophic Lateral Sclerosis, Dysphagia, Neurodegenerative disorders, Malnutrition, Pneumonia, Pain.

Introduction

The treatment of dysphagia in neurodegenerative diseases aims to prolong life expectancy and improve the quality of life. With these patients one of the major causes of death is aspiration pneumonia.[1-3]

Among the motor neuron diseases, Amyotrophic Lateral Sclerosis (ALS) is the most frequent.[1] It is a neurodegenerative disease with a life expectancy of three to five years after onset of the first symptoms.[4,5] Patients with ALS who have a bulbar involvement at the onset of the disease, showing symptoms such as dysphagia and dysarthria, have an even lower life expectancy.[6]

The ALS is characterized by motor paralysis due to degeneration of motor neurons in the primary motor cortex, brainstem and spinal cord, without impairment of sensitivity.

Dysphagia in ALS is progressive and has its origin in weakness and spasticity of the muscles innervated by the trigeminal nerve, facial, hypoglossal, glossopharyngeal and vagus.[2]

The identification of dysphagia severity, with its associated factors which lead to nutritional and pulmonary complications, is a form of prevention. Even today many of these patients do not receive the desired attention and treatment for dysphagia and is the eventual cause of death due to the consequences of this change in the swallowing mechanism.

Non-motor symptoms, as pain, in ALS are not frequently reported. But studies indicate that 50 to 70% of patients with bodily pains.[7,8] The most often reported pain location is in the upper and lower extremities, shoulders and hips.[9] There are no studies about odynophagia (painful swallowing) in ALS, nor about its relationship with dysphagia

severity. The pain as a symptom in ALS is understudied and is unknown by many professionals.

It is a muscular pain and can be secondary to spasticity, atrophy, stiffness and reduced muscle tone. Pain can cause mobility reduction, decreasing motor coordination and affecting muscular function.[10]

The purpose of this study was to identify associated factors with dysphagia severity in ALS.

Material and Method

Selection of Patients

It was a cross-sectional study conducted with 49 patients previously diagnosed with ALS (according to the *El Escorial* criteria)[11] in a large Brazilian university hospital. All patients answered a verbal questionnaire administered by a speech-language therapist about their swallowing complaints and underwent Fiberoptic Endoscopy Evaluation of Swallowing (FEES).

The study included patients with ALS who had one or more complaints of swallowing and who were being treated with *Riluzol*. We excluded the patients who for some reason could not accomplish the FEES, who had frontotemporal involvement or another disorder that could also lead to changes in swallowing.

Procedures

At the clinical evaluation the patients' current age, age at onset of ALS symptoms, complaints of oral and pharyngeal phase of swallowing were obtained.

Patients answered the verbal questionnaire with “yes” or “no” as to oral complaints (labial food escape, chewing difficulty, palatal food residues, difficulty to eject the bolus, residue of food in the oral cavity, labial escape of liquid or saliva and difficulty in swallowing saliva) and as to pharyngeal complaints (nasal reflux of food or liquid, coughing, gagging, throat clearing during or after feeding, sensation of food stasis, painful swallowing (odynophagia), swallowing difficulties and swallowing delays).

During the examination three types of food were offered: 1) Lemon juice colored with green dye. 2) Nectar, honey and pudding consistencies, all colored with green dye (these fluids were obtained with the addition of 2, 3 and 4 teaspoons of a thickener (Thicken-easy®) to 100 mL of water, respectively, and were offered in two different quantities, 3 mL and 7 mL). 3) A solid consistency was represented by a cornstarch biscuit.

The food was given to patients in the following sequence: liquid and nectar (3 mL, 3 mL, 7 mL and 7 mL); honey (3 mL, 3 mL, 7 mL and 7 mL); pudding (2 tablespoons); solid (½ cornstarch biscuit). The liquid food was administered in 20 mL syringes, with the sample introduced into the patient’s oral cavity. As difficulties in swallowing were observed, protective maneuvers of the airways and/or changes in head posture were performed in order to assist oral feeding in a safe way.

FEES’s were carried out by an otorhinolaryngologist while the food was offered by a speech-language therapist.

For analyses, the results of FEES were judged by two speech therapists and one otorhinolaryngologists. They were categorized according to the classification of Macedo Filho et al.[12]: Level 0) Normal swallowing: normal oral containment, presence of reflexes, absence of salivary or food stasis, less than three propulsion attempts to clear the bolus. Level 1) Mild Dysphagia: limited stasis after swallowing, less than three propulsion

attempts to clear the bolus, no nasal regurgitation or laryngeal penetration. Level 2) Moderate dysphagia: moderate salivary stasis, greater stasis post-swallowing, more than three attempts to clear the bolus, nasal regurgitation, reduced laryngeal sensitivity with penetration, but without laryngotracheal aspiration. Level 3) Severe dysphagia: large salivary stasis, marked impairment in the amount of residue after swallowing, weak or absent propulsion, nasal regurgitation, laryngotracheal aspiration.

Data Analysis

The patients were divided into groups according to the dysphagia severity classified by FEES. Group 1) patients with normal swallowing or mild dysphagia, Group 2) patients with moderate or severe dysphagia.

Descriptive analysis was performed for the frequency of categorical variables and median measures and standard deviation for numeric variables.

To identify associated factors with moderate or severe dysphagia Cox Regression was used to analyze with single to multiple variables, using a stepwise process. The multiple variable analyses were used to exclude those variables which did not apply and to obtain a factor that more closely associated with dysphagia severity.

The dysphagia severity (group 1 or 2) was considered the dependent variable in Cox Regression. The independent variables tested were: age at assessment, age at onset of first symptoms, disease duration (time estimated between the age at onset of symptoms and the age at assessment), gender, number and type of swallowing complaints of oral phase and number and type of swallowing complaints of pharyngeal phase.

The analyses were performed using Statistical Package for the Social Sciences (SPSS) version 13.0 for Windows and statistical tests considered p-values less than or equal to 0.05.

Results

Based on the classification of the dysphagia severity, patients were distributed in groups, with 25 allocated to Group 1 (three (6.1%) had normal swallowing and 22 (44.9%) had mild dysphagia) and 24 to Group 2 (20 (40.8%) had moderate dysphagia and 4 (8.1%) severe dysphagia).

Table 1 shows the descriptive analysis for the characterization of the patients. Tables 2 and 3 show the frequencies of swallowing complaints in the oral and pharyngeal phases respectively.

The number of swallowing complaints of the oral and pharyngeal phases, the reference of difficulty to eject the bolus and odynophagia were identified as associated factors with moderate or severe dysphagia, in single variable analysis (Table 4).

Only the odynophagia remained as an associated factor of moderate or severe dysphagia in multiple variable analysis (risk: 11.96; confidence interval: 1.60 to 89.36, p-value: 0.01). The odynophagia was observed by 31 (63.2%) patients, nine (36%) of group 1 and 22 (91.6%) of group 2.

Table 1. Descriptive analysis of patients with ALS classified according to dysphagia severity.

	Group 1: Normal swallowing and mild dysphagia (N=25)	Group 2: Moderate and severe dysphagia (N=24)
	Mean (± SD[a])	Mean (± SD[a])
Gender (male/female)	13/12	14/10
Age at symptoms' onset (years)	52.1 (± 10.5)	52.7 (± 9.6)
Age at first assessment (years)	54.4 (± 11)	55 (± 9.2)
Disease duration (years)	2.29 (± 1.8)	2.2 (±1.5)
Swallowing complaints (N)	5.6 (± 4)	10 (± 2)
Swallowing complaints of oral phase (N)	2.6 (± 2.5)	4.8 (± 2)
Swallowing complaints of pharyngeal phase (N)	3 (± 2)	5 (± 1.3)

[a] Standard Deviation (SD)

Table 2. Frequency of swallowing complaints in the **oral phase** observed by patients with ALS classified according to dysphagia severity.

Complaints	Group 1: Normal swallowing and mild dysphagia (N=25)	Group 2: Moderate and severe dysphagia (N=24)
	N (%)	N (%)
Labial food escape	10 (40.0)	16 (66.6)
Chewing difficulty	13 (52.0)	17 (70.8)
Palatal food residues	12 (48.0)	13 (54.1)
Difficulty to eject the bolus	10 (40.0)	20 (83.3)
Residue of food in the oral cavity	12 (48.0)	17 (70.8)
Labial escape of liquid or saliva	13 (52.0)	17 (70.8)
Difficulty in swallowing saliva	4 (16.0)	4 (16.6)

Table 3. Frequency of swallowing complaints in the **pharyngeal phase** observed by patients with ALS classified according to dysphagia severity.

Complaints	Group 1: Normal swallowing and mild dysphagia (N=25)	Group 2: Moderate and severe dysphagia (N=24)
	N (%)	N (%)
Nasal reflux	5 (20.0)	7 (29.1)
Coughing	4 (16.0)	4 (16.6)
Gagging	16 (64.0)	22 (91.6)
Throat clearing	14 (56.0)	16 (66.6)
Sensation of food stasis	19 (76.0)	16 (66.6)
Painful swallowing (odynophagia)	9 (36.0)	22 (91.6)
Swallowing difficulties	11 (44.0)	16 (66.6)
Swallowing delays	2 (8.0)	3 (12.5)

Table 4. Associated factors of moderate or severe dysphagia, identified by the single variable analysis of Cox Regression in a sample of 49 ALS Brazilian patients.

Variables	Prevalence	Confidence Interval	P-value
Swallowing complaints of oral phase (N)	1,38	1,08 – 1,75	0,009
Swallowing complaints of pharyngeal phase (N)	1,20	1,01 – 1,43	0,03
Difficulty to eject the bolus (yes x no)	3,16	1,08 - 9,26	0,03
Painful swallowing (odynophagia) (yes x no)	6,38	1,50 – 27,16	0,01

Discussion

Associated factors with dysphagia in ALS

Dysphagia in patients with ALS may be associated with atrophy and weakness of the tongue, caused by nuclear or supranuclear lesion of the hypoglossus, vagus and glossopharyngeal nerves, leading to failure in the closure of the soft palate, nasal reflux and change in larynx closure, reducing airway protection.[13]

The results of this study show practically all patients already suffering from some level of dysphagia at the time of evaluation. In ALS, generally all patients will suffer from dysphagia at some time during the course of the disease. It is a rapidly evolving disease and in this study the majority of the patients were diagnosed with mild or moderate dysphagia in little more than two years of ALS duration. Studies conducted in the United States and Italy found similar prevalence of mild and moderate dysphagia in ALS population.[1,14]

We chose to examine separately the complaints of the oral and pharyngeal phases of swallowing because the literature shows that dysphagia in ALS usually starts by dysfunctions in the oral phase of swallowing due to the difficulty in managing and transporting the bolus.[13,15] To Kawai et al.[13] the function of tongue ejection for managing and movement the bolus is possibly already affected in the initial stage of the disease. According to the literature, the problem is worsened in the pharyngeal phase, including reduced laryngeal elevation, decreased elevation of the soft palate and deficit in pharyngeal contraction.[16,17]

In the present study, patients with mild dysphagia or normal swallowing did not report more complaints in the oral phase than in the pharyngeal phase. They differed only as to the amount of complaints, and the individuals with moderate or severe dysphagia described a greater number of swallowing complaints.

In ALS due to the preservation of oropharyngeal sensitivity, the patient perceived each of their dysphagia symptoms and felt the increased frequency with which symptoms appeared in their feeding. The analysis showed that the greater the number of swallowing complaints, the greater the risk that the patient had already been affected by a moderate or severe dysphagia.

In the highest risk group, composed by individuals diagnosed with moderate or severe dysphagia, one of the most perceived swallowing complaints was throat clearing after or during feeding.

Throat clearing is a protective mechanism of the lower airway, suggesting failure during the swallowing act. The reduced ejection force, decreased pharyngeal peristalsis, reduced laryngeal elevation and the upper sphincter of the esophagus hypertonicity allow residues in the vallecula, posterior pharynx, pyriform sinuses and upper sphincter of the esophagus. The non-removal of such residues, also known as stasis, during swallowing, is primarily responsible for laryngotracheal aspiration in ALS patients.[18]

The lack of oral control of the bolus is a result of lingual paralysis. Furthermore, the weakness of the pharyngeal constrictor muscles delays the passage of the bolus and the ineffective closure of the larynx allows penetration of the bolus in the airways, causing coughing and throat clearing.[15]

Just as the throat clearing, coughing is a protective reflex of the lower airway, witch however, was not mentioned by most participants. Adequate cough strength is crucial for airway protection. In ALS patients, due to progressive weakness of the muscles of the larynx and breathing, cough becomes impaired.[19]

The difficulties in eject the bolus was often referred to by patients with high risk (group 2) and was associated with moderate or severe dysphagia. The weakness and muscle

paralysis also affected the efficiency of eject the bolus. In some instances dysphagia starts with difficulties in moving the bolus from the front of the tongue, it began with reduced ability to retain the bolus at the back of the tongue.[1] At a later phase, the patient with ALS notices dysphagia as a almost complete inability to propel the bolus to the pharynx, causing increased risk of laryngeal penetration and aspiration.

Therefore, it is important to consider the complaint of the patient in order to offer other options to compensate their swallowing impairment, enabling him to maintain a safe oral feeding whenever possible.

Odynophagia in ALS

There are controversies about pain in ALS. The results of this study confirmed with multiple variable analysis that there is odynophagia (painful swallowing) associated with dysphagia severity in ALS.

This study was limited by the extensive range of the confidence interval. This means that odynophagia can be identified as an associated factor with moderate or severe dysphagia, but the accuracy of this prediction, i.e. how its occurrence may increase the risk of moderate or severe dysphagia, should not be generalized.

In literature coughing is the major associated factor with the risk of laryngotracheal aspiration.[20] However, as discussed before, coughing in patients with ALS is usually weak or absent.

Despite the fact that many professionals consider ALS as a painless disease; this study presents new evidence demonstrating that these patients also report pain during swallowing in the more advanced stages of dysphagia.

The sensation of pain in ALS can be explained by the progressive degeneration of electrical and mechanical properties of the muscles, which causes reduction in strength and in coordination of the muscles, generating a kind of stress on the ligaments, tendons and joints. Such changes occur repeatedly and constantly and result in minor inflammation and consequent pain.[21]

The spasms, contractions and abnormal stress on the system, imposed by skeletal muscle weakness, may also cause pain due to immobilization of the articular blocks.[22]

As the disease progresses and mobility decreases, body pain becomes more frequent.[23] The disease, which leads to greater weakness and reduced mobility of oropharyngeal muscles, generates greater pain and impairment in the swallowing function. Therefore, the pain ends up being an associated factor of more severe stages of dysphagia.

The patient with ALS can reference symptoms of dysphagia not only by the frequency of gagging or coughing (symptoms more commonly associated with the dysphagia severity), but also, and maybe more specifically, by painful swallowing.

There is evidence that pain in ALS interferes in the activities of daily life, aside from interfering in their social lives.[24] However, in this disease, the pain is usually one of the less studied, most underrated and potentially least treated. It is a symptom rarely discussed and often not reported by patients.[25] A recent Dutch study refers to pain as a symptom unrecognized and untreated in ALS.[26] The authors emphasize that efforts need to be employed to identify it and treat it appropriately.

Besides affecting social life, the pain also influences feeding. The pain is not an emphasized aspect in the evaluation and swallowing treatment of ALS patients. However, the present study reveals that special attention should be given to this complaint as it may indicate a worsening of dysphagia. Being a progressive neurodegenerative disease with

short life expectancy, dysphagia is progressive and has a significant impact in these patients' quality and duration of life. Adequate monitoring of its evolution may indicate the need for other treatment, such as airway protection maneuvers, bolus consistency changes and even an indication of non-oral feeding.

More studies are necessary for odynophagia in ALS in order to have a better comprehension of this symptom and to investigate an effective management.

Conclusion

The most consistently associated factor with dysphagia severity in ALS was odynophagia. Its presence in ALS patients suggests a high risk of pulmonary and nutritional complications.

References

(1) Fattori B, Grosso M, Bongioanni P, et al. Assessment of swallowing by oropharyngoesophageal scintilography in patients with amyotrophic lateral sclerosis. Dysphagia 2006; 21:280-6.

(2) Janzen VD, Hae R, Hudson AJ. Otolaryngologic manifestations of amyotrophic lateral sclerosis. J Otolaryngol 1996; 17: 41–2.

(3) Hadjikoutis S, Wiles CM. Respiratory complications related to bulbar dysfunction in motor neuron disease. Acta Neurol Scand 2001; 103:207-13.

(4) Oliveira ASB, Pereira RDB. Amyotrophic Lateral Sclerosis (ALS): three letters that change the people's life. Arq Neuropsiquiatr 2009; 67(3-A):750-82.

(5) O'toole O, Traynor BJ, Brennan P et al. Epidemiology and clinical features of amyotrophic lateral sclerosis in Ireland between 1995 and 2004. J. Neurol Neurosurg Psychiatry 2008; 79:30-2.

(6) Zoccolella S, Beghi E, Palagano G, Fraddosio A, Guerra V, Samarelli et al. Analysis of survival and prognostic factors in amyotrophic lateral sclerosis: a population based study. J Neurol Neurosurg Psychiatry 2008; 79:33–37.

(7) Ganzini L, Johnston WS, Hoffman WF Correlates of suffering in amyotrophic lateral sclerosis. Neurology 1999; 52:1434–40.

(8) Oliver D. The quality of care and symptom control—the effects on the terminal phase of ALS/MND. J Neurol Sci 1996; 139(Suppl):134–136.

(9) Chiò A, Canosa A, Gallo S, Moglia C, Ilardi A, Cammarosano S. et al. Pain in amyotrophic lateral sclerosis: a population-based controlled study. Eur J Neurol 2012; 19:551-55.

(10) Thomas CK, Zijdewind I. Fatigue of muscles weakened by death of motoneurons. Muscle and Nerve 2006; 33(1):21–41.

(11) Brooks BR. El Escorial World Federation of Neurology criteria for the diagnosis of amyotrophic lateral sclerosis. Subcommittee on Motor Neuron Diseases/Amyotrophic Lateral Sclerosis of the World Federation of Neurology Research Group on Neuromuscular Diseases and the El Escorial "Clinical limits of amyotrophic lateral sclerosis" workshop contributors. J Neurol Sci. 1994 Jul;124 Suppl:96-107.

(12) Macedo Filho ED, Gomes GF, Furkim AM. Manual de cuidados do paciente com disfagia. São Paulo: Lovise, 2000.

(13) Kawai S, Tsukuda M, Mochimatsu I, Enomoto H, Kagesato Y, Hirose H et al. A study of the early stage of dysphagia in amyotrophic lateral sclerosis. Dysphagia 2003; 18:1–8.

(14) Leder SB, Novella S, Patwa H. Use of Fiberoptic Endoscopic Evaluation of Swallowing (FEES) in Patients with Amyotrophic Lateral Sclerosis. Dysphagia 2004; 19: 177-81.

(15) Pontes RT, Orsini M, Freitas MRG, Antonioli RS, Nascimento OFM. Alterações da fonação e deglutição na Esclerose Lateral Amiotrófica: Revisão de Literatura. Rev Neurocienc 2010; 18(1):69-73.

(16) Ohkubo H. Dysphagia in Amyotrophic Lateral Sclerosis – Eletromiotrofic and radiological interventions. Otol Fukuoka 1980; 26:44-88.

(17) Okamura H, Mori T, Inake S. Dysphagia due to neuromuscular diseases. J Jpn Bronchoesophagol Soc 1991; 42:400-06.

(18) Yokoyama M, Mitomi N, Tetsuka K, Tayama N, Niimi S. Role of laryngeal movement and effect of aging on swallowing pressure in the pharynx and upper esophageal sphincter. Laryngoscope 2000; 110:434–9.

(19) Mari F, Matei M, Ceravolo MG, Pisani A, Montesi A, Provinciali L et al. Predictive value of clinical indices in detecting aspiration in patients with neurological disorders. J Neurol Neurosurg Psychiatry 1997; 63: 456-60.

(20) Garon BR, Engle M, Ormiston C. Reliability of the 3-oz water swallow test utilizing cough reflex as sole indicator of aspiration. J Neurol Rehab 1995;9:139–43.

(21) Handy CR, Krudy C, Boulis N, Federici T. Pain in Amyotrophic Lateral Sclerosis: A Neglected Aspect of Disease. Neurol Res Int 2011; 1:1-8.

(22) Brettschneider J, Kurent J, Ludolph A, Mitchell JD. Drug therapy for pain in amyotrophic lateral sclerosis or motor neuron disease. Cochrane Database Syst Rev 2008; 3:CD005226.

(23) Simmons Z. Management strategies for patients with amyotrophic lateral sclerosis from diagnosis through death. Neurologist 2005; 11:257–70.

(24) Pagnini F, Lunetta C, Banfi P, Rossi G, Fossati F, Marconi A et al. Pain in Amyotrophic Lateral Sclerosis: a psychological perspective. Neurol Sci 2011 [epub ahead of print].

(25) Miller RG, Anderson FA Jr, Bradley WG, Brooks, BR; Mitsumoto, H; Munsat et al. The ALS patient care database: goals, design, and early results. ALS C.A.R.E. Study Group. Neurology 2000; 54: 53–57.

(26) Maessen M, Veldink JH, van den Berg LH, Schouten HJ, van der Wal Onwuteaka-Philipsen BD. Requests for euthanasia: origin of suffering in ALS, heart failure, and cancer patients. J Neurol 2010; 257: 1192–8.

CAPÍTULO 2

AMYOTROPHIC LATERAL SCLEROSIS: SURVIVAL ANALYSIS OF SWALLOWING AND NON-ORAL FEEDING

Esclerose Lateral Amiotrófica: Análise de sobrevivência da deglutição e da alimentação por via alternativa

Autores: Luchesi KF, Kitamura S, Mourão LF.

(Submetido)

Abstract

Aim: To analyze aspects associated with worsened swallowing functionality and need for non-oral feeding in Amyotrophic Lateral Sclerosis (ALS) patients.

Methods: This is a long-term study of 33 ALS patients (28 with spinal onset ALS and five with bulbar onset ALS). The patients were observed between 2006 and 2011 at intervals of three months. They underwent Fiberoptic Endoscopic Evaluation of Swallowing (FEES) and Functional Oral Intake Scale (FOIS) classification. In order to study the factors related to the time prior to the onset of worsened swallowing and prior to the indication of non-oral feeding, Survival Analysis was performed using Cox Regression.

Results: There was incidence of worsened swallowing functionality and indication of non-oral feeding in 84.8% and 36.4% of the patients respectively. The worsening of swallowing functionality was more rapid in individuals who were older at the onset of ALS (increased risk of 1.03 (3%) per year) and in individuals with bulbar onset ALS (increased risk of 2.88 times). The non-oral feeding indication process was associated with patients who were older at the onset of ALS symptoms (increased risk of 1.06 (6%) per year) and shorter disease duration (increased risk of 1.096 (9.6%) per year).

Conclusions: In these patients, the worsened swallowing was faster in individuals who were older at the onset of symptoms and who had bulbar onset ALS. The non-oral feeding indication process was associated with patients who were older at the onset of ALS symptoms or had the disease a shorter time.

Key Words: dysphagia; deglutition; deglutition disorders; Amyotrophic Lateral Sclerosis; disease progression.

Introduction

Dysphagia is a disorder that compromises one or more stages of swallowing. It is usually a symptom of some underlying disease and manifests itself as coughing and choking during or after a meal, chewing difficulty, drooling, weight loss and aspiration pneumonia [1]. It is a frequent counterpart of neurological disorders, especially those with prominent motor dysfunction.

Amyotrophic Lateral Sclerosis (ALS) is a devastating neurodegenerative disease of the motor pathways [2, 3]. It is the most frequent motor neuron disease in adults, with a worldwide incidence from 1 to 3/100.000 people per year [4, 5]. It is characterized by the progressive degeneration of lower and upper motor neurons in the cerebral cortex, brainstem, and spinal cord, resulting in muscular atrophy, fasciculation, weakness and spasticity [2]. The survival period of ALS patients ranges from three to five years [2, 3].

The onset of signs and symptoms of ALS are used to classify the disease in spinal and bulbar onset. In bulbar onset, ALS is first observed involving the tongue muscles resulting in dysphagia and dysarthria as initial symptoms [6]. In spinal onset, ALS is first observed involving the muscles of extremist resulting in weakness and/or reduction of mobility.

Dysphagia in ALS patients is progressive and is due to weakness or spasticity of muscles innervated by the trigeminal, facial, hypoglossal, glossopharyngeal or vagal nerves [7]. Dysphagia-related complications, such as aspiration pneumonia and malnutrition are major causes of death among patients with ALS [4, 8]. Dysphagia is one of the most important symptoms in the prognosis of ALS and correct diagnosis. Appropriate therapeutic intervention and timely follow-up of swallowing are highly necessary [9].

There is evidence that multidisciplinary palliative approach can prolong survival and maintain the quality of life in ALS patients [2]. Those treated in the early stages of the disease can develop the adaptation of muscle mechanisms to decrease broncoaspiration risk [10].

There are few studies outlining the severity and temporal course of dysphagia progression in ALS patients [11]. Familiarity with the factors influencing swallowing functionality can enable speech-language therapists and neurologists to design more focused treatment strategies and prevent some of the complications associated with the disease. One of these complications is the maintenance of oral feeding when the patient lacks the ability to swallow safely. A delay in the introduction of non-oral feeding can avoid complications such as the previously mentioned aspiration pneumonia, malnutrition and, ultimately, death.

This article aims to analyze the aspects associated with worsened swallowing functionality and the need for non-oral feeding for ALS patients.

Material & methods

Selection of Patients

This was a prospective and retrospective long-term study of 33 dysphagic patients (28 with spinal onset ALS, five with bulbar onset ALS), observed in a five year period (2006-2011) in a large Brazilian university hospital. It was an open case series and during the five years of study there were patients moving in and out.

Prior to participating in the study, ALS was diagnosed in the patients according to the modified *El Escorial* [12] criteria. The patients were observed and regularly followed-up throughout the course of their outpatient swallowing treatment.

Only patients who had at least two evaluations, complaints of swallowing, no prior non-oral feeding and regular check-ups with Outpatient Neurology were included in the study. Patients with frontotemporal involvement or concomitant disorders able to cause dysphagia were excluded from the analysis.

Procedures

The patients underwent evaluations for swallowing and Functional Oral Intake Scale (FOIS) every three months.

The swallowing evaluation, upon which non-oral feeding indication was based, was obtained through a Fiberoptic Endoscopic Evaluation of Swallowing (FEES) and clinical evaluation. In both evaluations, three types of food were offered: 1) Lemon juice colored with green dye. 2) Nectar, honey and pudding consistencies, all colored with green dye (these fluids were obtained with the addition of 2, 3 and 4 teaspoons of a thickener (Thicken-easy®) to 100 mL of water, respectively, and were offered in two different quantities, 3 mL and 7 mL). 3) A solid consistency was represented by a cornstarch biscuit.

The food was given to patients in the following sequence: liquid and nectar (3 mL, 3 mL, 7 mL and 7 mL); honey (3 mL, 3 mL, 7 mL and 7 mL); pudding (2 tablespoons); solid (½ cornstarch biscuit). The liquid food was administered in 20 mL syringes, with the sample introduced into the patient's oral cavity. As difficulties in swallowing were observed, protective maneuvers for the airways and/or changes in head posture were performed in order to assist oral feeding in a safe way.

FEES were carried out by an otorhinolaryngologist while the food was offered by a speech-language therapist.

In the clinical evaluation, as the patients swallowed, cervical auscultation was performed to identify abnormal signs in the pharyngeal swallowing phase. Oral bolus transit time, anterior or posterior escape, positive cervical auscultation (with signs that indicate a presence of stasis or penetration with aspiration risk), coughing (before, during or after swallowing) and wet voice were also observed.

Based on the clinical evaluation and FEES the Functional Oral Intake Scale (FOIS) [13] was applied. Its purpose was to classify the swallowing functionality of all patients every three months. The FOIS ranks patients into levels. For the present study the levels are described as follows: Level 1) Nothing by mouth. Level 2) Tube dependent with minimal attempts of food or liquid. Level 3) Tube dependent with consistent oral intake of food or liquid. Level 4) Total oral diet of one or two consistencies (nectar and honey, honey and pudding). Level 5a) Total oral diet with multiple consistencies but with restriction of two consistencies (for example, solid and liquid), with or without compensation. Level 5b) Total oral diet with multiple consistencies but with restriction of one consistency (for example, solid or liquid), with or without compensation. Level 5c) Total oral diet with multiple consistencies, but requiring compensation. Level 6) Total oral diet with multiple consistencies without special preparation but with specific food limitations (for example: fibers, grains and some vegetables) and speed and volume modification if necessary. Level 7) Total oral diet with no restriction.

In FOIS, a patient's classification into levels 1, 2 or 3 means that their swallowing functionally and/or their clinical conditions does not allow a sufficiently healthy oral diet and requires non-oral supplementation.

At the initial evaluation, every patient received a corresponding FOIS classification. Every time the patients returned to the hospital they were reevaluated and reclassified with

FOIS. To analyze the worsened swallowing functionality, patients were observed until the point of the time at which a fall in FOIS level or sublevel was noted. To analyze the need for non-oral feeding, patients were observed until the point of time at which they required tube feeding and were classified at FOIS level three.

This article does not aim to discuss the results of FEES; it was described only to clarify for the readers the process of swallowing evaluation, upon which non-oral feeding indication was based.

Data Analysis

In order to study the factors related to the time prior to the onset of worsened swallowing functionality and the necessity of non-oral feeding, Survival Analysis was performed using Cox Regression.

The time from the onset of the ALS symptoms until the event (the worsened swallowing and/or the indication of non-oral feeding) was considered the dependent variable. Age at the first evaluation at the Outpatient Clinic, age at the onset of symptoms, disease duration (time from the onset of symptoms to the first visit to the clinic), ALS onset and gender were considered as independent variables.

Analysis was performed using the software Statistical Package for the Social Sciences (SPSS) version 13.0 for Windows, and p-values lower than 0.05 were considered significant.

It is important to mention that the results of a survival analysis express the probability of the patient not suffering from a given event over time.

This study was approved by the Ethic Board Committee of the Faculty of Medical Sciences at the University of Campinas, Brazil (Protocol Number 796/2005).

Results

There was incidence of worsened swallowing functionality for 84.8% (28 patients) and incidence of indication of non-oral feeding for 36.4% (12 patients). Table 1 shows a descriptive analysis.

In the twentieth month of observation, Survival Function showed a 60% probability of not experience worsened swallowing in spinal onset. This probability dropped to 40% in bulbar onset ALS (Figure 1). In the same period of observation, Hazard Function showed a 50% probability of not needing non-oral feeding in spinal onset ALS. The probability dropped to 15% in bulbar onset ALS (Figure 2).

The older patient was at the onset of ALS symptoms increased the risk by 4% (risk: 1.04) per year according to Cox Regression multiple variable analysis. The patients with bulbar onset ALS had a 2.88 increased risk of worsening the swallowing functionality in a short period.

Concerning the factors related to necessity of non-oral feeding, women were 3.47 times more likely to be at risk. However, in the multiple variable analysis, only the patient's age at the onset of symptoms and the disease duration was significant. The age at the onset of symptoms increases in 7.5% per year the risk of non-oral feeding necessity (risk: 1.075). Disease duration was inversely related to the time of non-oral feeding process indication. Shorter disease duration showed an increased risk of 13% per year (risk: 1.13) to start non-oral feeding (Table 3).

Table 1. Descriptive analysis of variables: gender, age at first evaluation, age at onset of symptoms, disease duration and time prior to: worsened swallowing functionality and indication of non-oral feeding in a case series of ALS Brazilian patients followed between 2006 and 2011 (N=33).

	Bulbar onset ALS (N=5)			**Spinal onset ALS (N=28)**		
Variables	*Min-Max*	*Mean±SD*	*Median*	*Min-Max*	*Mean±SD*	*Median*
Gender (male/female)		2/3			16/12	
Incidence of worsened swallowing		100%(5)			82.1%(23)	
Incidence of non-oral feeding indication		40%(2)			35.7%(10)	
Age at first clinic visit (years)	50-66	56±6	55	37-73	55.2±10.3	57
Age at onset of symptoms (years)	50-66	54.4±6.5	54	36-71	52.7±10.5	54
Disease duration (months)	12-36	19.2±10.7	12	12-96	30.4±21.2	24
Time up to worsened swallowing functionality (months)*	13-48	24.6±14.2	18	7-105	36.5±25.7	24
Time up to non-oral feeding indication (months)*	13-58	28.8±19.3	18	14-116	48±27.3	40.5

*Time estimated from the onset of ALS symptoms.

Table 2. Significant variables detected by survival analysis using Cox Regression, considering the **worsened swallowing functionality** in a case series of ALS Brazilian patient followed in a dysphagia outpatient between 2006 and 2011 (N=33).

Variables	Risk	Confidence Interval	P-value
Single Variable Analysis			
Age at the first evaluation	1.03	1.001 – 1.073	0.04
Age at symptoms' onset	1.04	1.005 – 1.076	0.02
Bulbar onset ALS	2.88	1.019 – 8.179	0.046
Multiple Variable Analysis			
Age at symptoms' onset	1.04	1.005 – 1.076	0.02
Bulbar onset ALS	2.88	1.019 – 8.179	0.046

Table 3. Significant variables detected by survival analysis using Cox Regression, considering the indication of **non-oral feeding** in a case series of ALS Brazilian patient followed in a dysphagia outpatient between 2006 and 2011 (N=33).

Variables	Risk	Confidence Interval	P-value
Single Variable Analysis			
Gender (female)	3.47	1.039 – 11.629	0.04
Age at symptoms' onset	1.06	1.006 – 1.134	0.03
Short disease duration	1.096	1.030 – 1.168	0.004
Multiple Variable Analysis			
Age at symptoms' onset	1.075	1.012 – 1.142	0.01
Short disease duration	1.13	1.034 – 1.240	0.07

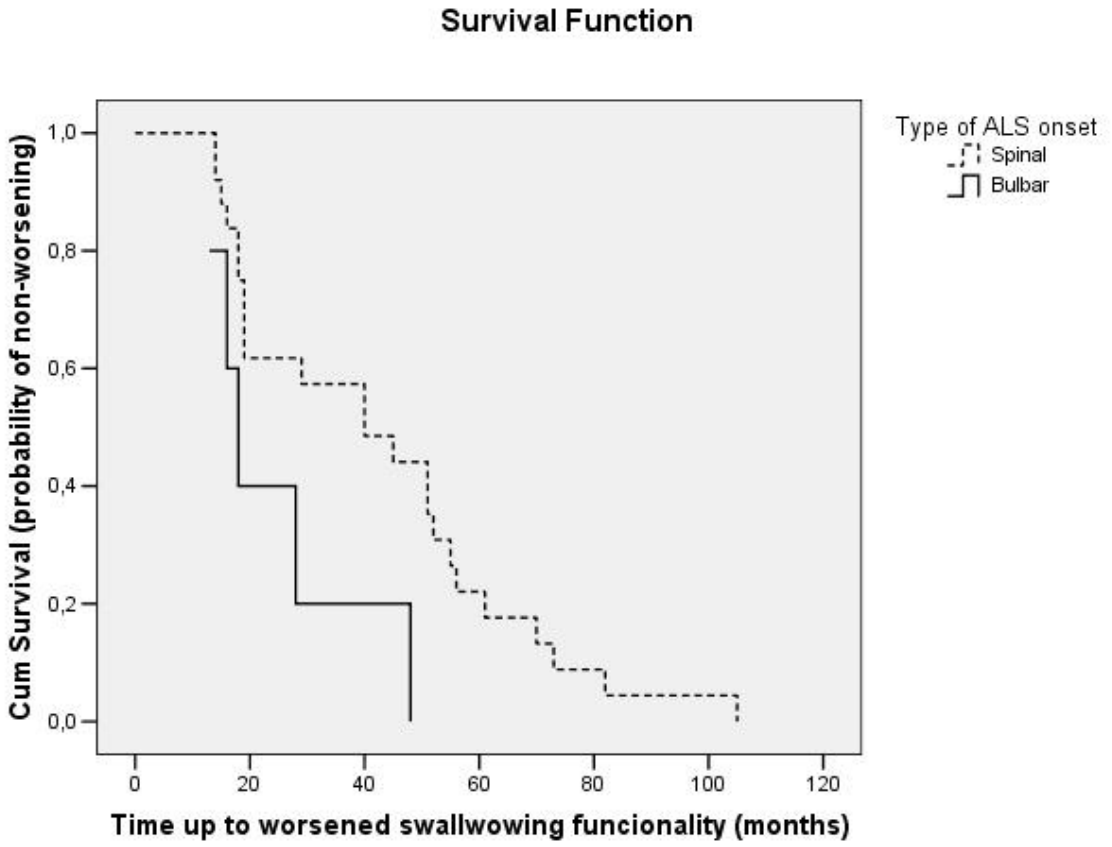


Figure 1. Survival plot considering **worsened swallowing functionality** of patients followed at a dysphagia outpatient between 2006 and 2011 (N=33). *Censored observation shows patients lost in follow-up or patients without worsened swallowing functionality during the observation period.

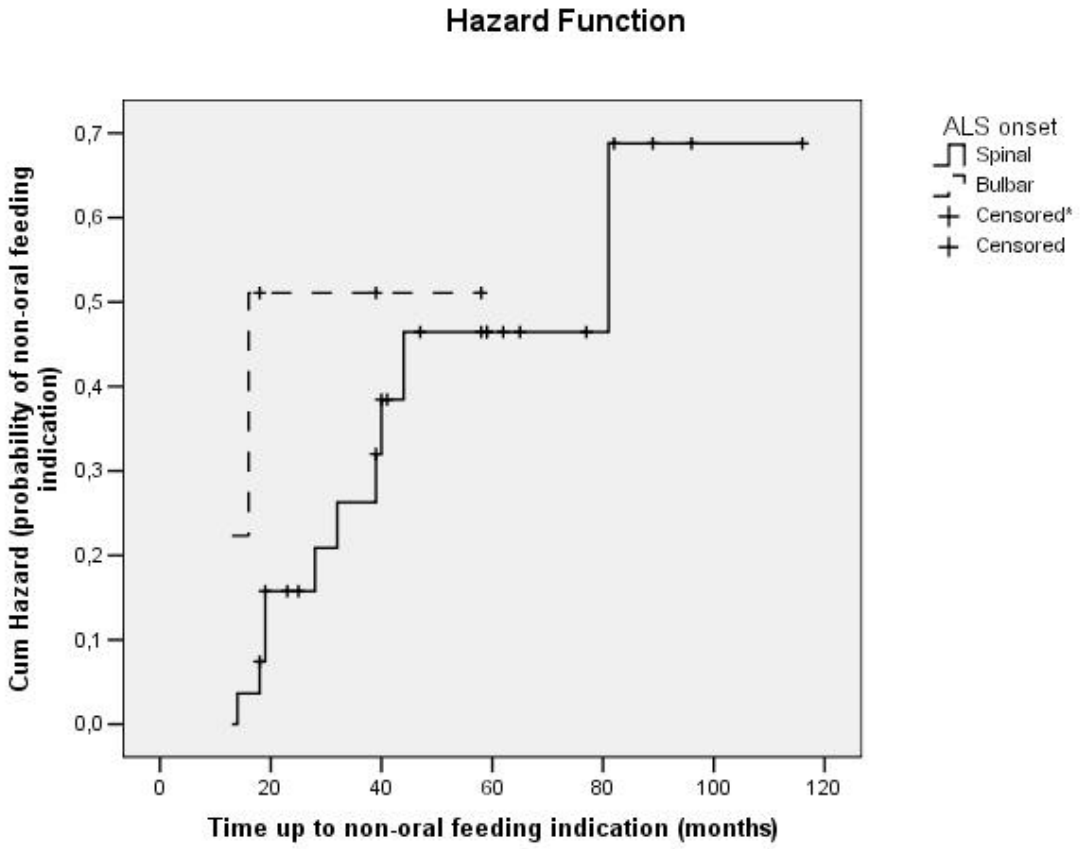


Figure 2. Survival plot considering the indication of **non-oral feeding** in patients followed at a dysphagia outpatient between 2006 and 2011 (N=33). *Censored observation shows patients lost in follow-up or patients without indication of non-oral feeding during the observation period.

Discussion

In this study, the number of men and women were similar. Recent studies suggest that the ALS prevalence tends to be almost equal with regards to gender [3, 16, 17]. Other studies indicate a slightly greater prevalence of ALS in men, with a ratio of 1.5:1 [18].

Our results show that the women tend to have worse swallowing functionality, with the necessity for non-oral feeding occurring before the men. Generally women and patients with bulbar onset ALS tend to have a worse prognosis in ALS [19]. According to the literature, men's tongue function has a significantly higher maximum pressure capability than women [20] and tongue pressure is very important to swallowing. There are findings that suggest women's glossopharyngeal muscle is more vulnerable in neurodegenerative diseases than men's [21]. Some authors propose that the possible reasons for the differences between genders in ALS include possible differences between male and female nervous systems, and different damage repairing abilities [22].

Studies report the age at which ALS appears ranges from 55 to 65 years old, with a median of 64 years [23, 24]. In relation to Brazilian citizens, the average age of ALS onset is around 52 years old [25]. One study performed in southern Brazil, which included 251 cases of ALS, referred to a patient age at the first evaluation of 54.4 (±12.3), and symptoms beginning 17.9 (±15.7) months prior to this first evaluation [26]. In the present study, patients had a median age of 53 years at the ALS onset and an average age at the initial evaluation of 55.3 years. That means the patients took more than two years to seek out the swallowing management program. This delay in treatment may be due to various reasons, including difficulties in accessing health care, lack of information and the latency between the onset of ALS symptoms and the first swallowing complaints. Time is crucial for ALS

patients, since they have a life expectancy of only three to five years [2, 3] and dysphagia always occurs, although the time of its manifestation differs [27].

The period between ALS onset and the beginning of swallowing management can be a risk factor for malnutrition, which, when associated with other factors, increases the risk of death in ALS patients by almost eight times [28]. Patients treated by multidisciplinary clinics seem to survive up to 7.5 months longer than patients who do not receive follow-up treatment at such clinics [29].

This study found a proportional relation between the age at onset of ALS symptoms and the time of worsened swallowing functionality. The later the symptoms began, the faster swallowing functionality worsened. This result corroborates studies that report a negative prognosis in older patients and those with other types of factors, such as ALS onset (worse in bulbar onset), progression rate of respiratory, bulbar and lower limb symptoms [3]. In reference to swallowing conditions, younger ALS patients' tongues seem to have significantly higher maximum pressure capabilities when compared to older patients [21].

Despite the ALS onset (bulbar or spinal), at the initial evaluation all patients already had bulbar involvement due to swallowing complaints. According to some authors, in ALS most patients experience difficulty swallowing during the course of the disease, with progressive worsening. Almost all patients with spinal onset demonstrate bulbar involvement at later stages of the disease [2].

In this article, the influence of disease duration in the need for non-oral feeding can be associated with the time of bulbar functions involvement. Actually, the disease duration can show the beginning of the bulbar symptoms. The patients had a faster bulbar

involvement, resulting in earlier swallowing complaints and faster impairment of the bulbar functions, requiring earlier non-oral feeding [9].

There is evidence that patients with bulbar onset ALS had faster disease progression and shorter survival periods in comparison to patients with spinal onset ALS [30]. As in this study, patients with bulbar onset ALS represent the minority. The literature shows prevalence around 30% of bulbar onset ALS in comparison with spinal onset ALS [31].

The faster worsening of swallowing in bulbar onset ALS indicated a need for frequent care in swallowing management. Survival is worse in patients with bulbar onset ALS and the dysphagia complications are the main cause of death [16]. It is necessary to maintain safe oral feeding for as long as possible, but not unduly postpone non-oral feeding.

The literature indicates that compensatory methods and dietary modifications should be used to ensure oral intake for as long as possible and to prevent food aspiration [32].

Conclusion

In these patients, worsened swallowing was more rapid in individuals who were older and whose the onset of ALS was bulbar. The need for non-oral feeding was associated with patients who were older at the onset of ALS symptoms or had the disease a shorter time.

References

[1] Groher ME, Crary MA. Dysphagia: clinical management in adults and children. St Louis: Mosby, 2009.

[2] Oliveira ASB, Pereira RDB. Amyotrophic Lateral Sclerosis (ALS): three letters that change the people's life. Arq Neuropsiquiatr 2009; 67(3-A):750-82.

[3] Logroscino G, Traynor BJ, Hardiman O et al. Descriptive epidemiology of amyotrophic lateral sclerosis: new evidence and unsolved issues. J Neurol Neurosurg Psychiatry 2008; 79:6-11.

[4] Fattori B, Grosso M, Bongioanni P, Nacci A et al. Assessment of swallowing by oropharyngoesophageal scintilography in patients with amyotrophic lateral sclerosis. Dysphagia 2006; 21:280-6.

[5] O'toole O, Traynor BJ, Brennan P et al. Epidemiology and clinical features of amyotrophic lateral sclerosis in Ireland between 1995 and 2004. J. Neurol Neurosurg Psychiatry 2008; 79:30-2.

[6] Langmore SE, Lehman ME. Physiologic deficits in the orofacial system underlying dysarthria in amyotrophic lateral sclerosis. J Speech Hear Res 1994; 37:28–37.

[7] Janzen VD, Hae R, Hudson AJ. Otolaryngologic manifestations of amyotrophic lateral sclerosis. J Otolaryngol 1996; 17: 41–2.

[8] Hadjikoutis S, Wiles CM. Respiratory complications related to bulbar dysfunction in motor neuron disease. Acta Neurol Scand 2001; 103:207-13.

[9] Leder SB, Novella S, Patwa H. Use of Fiberoptic Endoscopic Evaluation of swallowing (FEES) in patients with Amyotrophic Lateral Sclerosis. Dysphagia 2004; 19:177–81.

[10] Pontes RT, Orsini M, De Freitas MRG, Antonioli RS, Nascimento OJM. Speech and swallowing disorders in Amyotrophic Lateral Sclerosis: Literature Review. Rev Neurocienc 2010; 18(1):69-73.

[11] Sancho PO, Boisson D. What are management practices for speech therapy in amyotrophic lateral sclerosis? Rev Neurol (Paris) 2006; 162(2):273-4.

[12] Brooks BR. El Escorial World Federation of Neurology criteria for the diagnosis of amyotrophic lateral sclerosis. Subcommittee on Motor Neuron Diseases/Amyotrophic Lateral Sclerosis of the World Federation of Neurology Research Group on Neuromuscular Diseases and the El Escorial "Clinical limits of amyotrophic lateral sclerosis" workshop contributors. J Neurol Sci. 1994 Jul;124 Suppl:96-107.

[13] Crary MA, Mann GDC, Groher ME. Initial pychometric assessment of a functional oral intake scale for dysphagia in stroke patients. Arch Phys Med Rehabil 2005; 86:1516-20.

[14] Robbins J, Butler SG, Daniels SK, Gross RD, Langmore S, Lazarus CL, Martin-Harris B, McCabe D, Musson N, Rosenbek JC. Swallowing and dysphagia rehabiliation: Translating principles of neural plasticity into clinically oriented evidence. Journal of Speech, Language, and Hearing Research 2008; 51:S276–S300.

[15] Abhinav K, Stanton B, Johnston C et al. Amyotrophic Lateral Sclerosis in South-East England: a population-based study. The South-East England register for amyotrophic lateral sclerosis. Neuroepidemiology 2007; 29:44-8.

[16] Zoccolella S, Beghi E, Palagano G et al. Analysis of survival and prognostic factors in amyotrophic lateral sclerosis: a population based study. J Neurol Neurosurg Psychiatry 2008; 79:33-7.

[17] Armon C. Epidemiology of ALS/MND. In: Shaw P and Strong M, eds. Motor Neuron Disorders. Elsevier Sciences 2003; 167-206.

[18] Youmans SR, Stierwalt JAG. Measures of Tongue Function Related to Normal Swallowing. Dysphagia 2006: 102–11.

[19] Leigh PN. Amyotrophic lateral sclerosis. In Motor Neuron Disordersand related diseases. Edited by: Eisen AA, Sham PJ. Amsterdam: Elsevier; 2007. p 249-68. [Aminoff MJ, Boller F, Swaab DF (Series Editor): Handbook of Clinical Neurology].

[20] Traynor BJ, Codd MB, Corr B, Forde C, Frost E, Hardiman OM. Clinical features of amyotrophic lateral sclerosis according to the El Escorial and Airlie House diagnostic criteria. Arch Neurol 2000; 57:1171-6.

[21] McCombe PA, Henderson RD. Effects of gender in Amyotrophic lateral sclerosis. Gender Medicine 2010; 7(6): 557-67.

[22] Manjaly ZR, Scott KM, Abihnav K et al. The sex ratio in Amiotrofic Lateral Sclerosis: a population based study. Amiotroph Lateral Scler 2010; 11:439-42.

[23] Haverkamp LJ, Appel V, Appel SH: Natural history of amyotrophic lateral sclerosis in a database population. Validation of a scoring system and a model for survival prediction. Brain 1995; 118(3):707-19.

[24] Palermo S, Lima JMB, Alvarenga RP. Epidemiologia da Esclerose Lateral Amiotrófica - Europa/América do Norte/América Do Sul/Ásia: discrepâncias e similaridades. Rev Bras Neurol 2009; 45(2):5-10.

[25] Werneck LC, Bezerra R, Neto OS, Scola RH. A clinical epidemiological study of 251 cases of amyotrophic lateral sclerosis in the south of Brazil. Arq Neuro-Psiquiatr 2007; 65(2):189-95.

[26] Kawai S, Tsukuda M, Moshimatsu I et al. A study of early stage of dysphagia in Amiotrophic Lateral Sclerosis. Dysphagia 2003; 18:1-8.

[27] Desport JC, Preux PM, Truong TC, Vallat JM, Sautereau D, Couratier P. Nutritional status is a prognostic factor for survival in ALS patients. Neurology 1999; 53:1059-63.

[28] Traynor BJ, Alexander M, Corr B, Frost E, Hardiman O. Effect of a multidisciplinary amyotrophic lateral sclerosis (ALS) clinic on ALS survival: a population based study,1996-2000. J Neurol Neurosurg Psychiatry 2003; 74(9):1258-61.

[29] Higo R, Tayama N, Nito T. Longitudinal analysis of progression of dysphagia in Amyotrophic Lateral Sclerosis. Auris Nasus Larynx 2004; 31:247-54.

[30] Magnus T, Beck M, Giess R, Puls I, Naumann M, Toyka KV. Disease progression in amyotrophic lateral sclerosis: predictors of survival. Muscle Nerve. 2002; 25(5):709-14.

[31] Haverkamp LJ, Appel V, Apeel SH. Natural history of Amyotrophic Lateral Sclerosis in a database population: validation of a scoring system and a model for survival prediction. Brain 1995; 118(3):707-19.

[32] Ludolph AC, Prosiegel M, Riecker A. Diagnosis and treatment of bulbar symptoms in amyotrophic lateral sclerosis. Nat Clin Pract Neurol 2008; 4(7):366-74.

CAPÍTULO 3

DYSPHAGIA PROGRESSION AND SWALLOWING MANAGEMENT IN PARKINSON'S DISEASE: EXPLORATORY STUDY

Progressão e tratamento da disfagia na Doença de Parkinson: Estudo exploratório

Autores: Luchesi KF, Kitamura S, Mourão LF.

(Submetido)

Abstract

Purpose: This article aims to describe a swallowing management and investigate associated factors with swallowing functionality in a case series of patients with Parkinson's Disease (PD). Methods: It is a long-term study with 24 patients with PD. The patients were observed in a five year period (2006-2011). They underwent Fiberoptic Endoscopic Evaluation of Swallowing (FEES), Functional Oral Intake Scale (FOIS) and therapeutic intervention every three months. In the therapeutic intervention they received orientation about exercises to improve swallowing functionality. In order to investigate factors related to swallowing functionality, the Chi-square, Kruskal-Wallis and Fisher tests were used. The period of time for improvement or worsening of swallowing functionality was described by Kaplan-Meier Survival Analysis. Results: During the follow-up, ten patients improved, five stayed the same and nine worsened their swallowing functionality. The median time to improvement was ten months. Prior the worsening there was a median time of 33 months of follow-up. There was no associated factor with improvement or worsening of swallowing. The maneuvers frequently indicated in therapeutic intervention were: chin-tuck, bolus consistency, bolus effect, strengthening–tongue, multiple swallows and vocal exercises. Conclusion: There was found a predominance of improvement and maintenance of swallowing during the follow-up. There was no associated factor with swallowing functionality. The swallowing management was characterized by compensatory maneuvers and motor exercises with and without the act of swallowing.

Keywords: deglutition; deglutition disorders; dysphagia; Parkinson's Disease; disease progression; speech therapy.

Introduction

Dysphagia is common in individuals with neurological disorders and affects food intake which may lead to complications such as choking, malnutrition and pulmonary aspiration.[1]

Parkinson's Disease (PD) is one of the most common neurodegenerative diseases in an elderly population with a worldwide incidence between 1 to 20 per 1000 people/year.[2,3] It is characterized by impairment of basal ganglia in voluntary movements, causing resting tremor, rigidity, akinesia (or bradykinesia) and postural instability.[2,4]

Dysphagia in PD is very common and affects more than 80% of individuals, reflecting the underlying motor impairments and the extent of the disease's progression.[5] The swallowing difficulties most frequently associated with PD is related to the oral and pharyngeal phase, resulting in abnormal bolus formation, multiple tongue elevations, delayed swallowing reflex and prolongation of the pharyngeal transit time with repetitive swallows to clear the throat.[6]

These dysphagia-related impairments have a direct influence on the nutritional and health status of the patients and are associated with increased morbidity and mortality.[7,8] However, few studies describe the progression of dysphagia and its severity in PD.[9] There is very little information about the temporal aspect of dysphagia progression in PD.

The knowledge of the dysphagia progression in PD could decrease the risk of aspiration pneumonia, consequently decreasing the risk of death, since it is one of the most frequent causes of death in aspiration pneumonia.[10,11]

Swallowing management, through the utilization of methods that compensate for the alterations in the swallowing process, aims to preserve a safe oral feeding as long as possible. The knowledge of dysphagia progression in PD can orient physicians and

therapists on what to expect of their patients and what treatment may be necessary over time.

The maneuvers used in swallowing managements can be categorized by their objectives and characteristics. Robbins et al.[12] describe the first category as "compensatory", with the maneuvers of chin-tuck, head rotation, head tilt, head back, side-lying, bolus consistency and breath hold. The "motor without swallow" category includes range of motion, strengthening–tongue, strengthening–respiratory, tongue control, Shaker, Lee Silverman Voice Treatment (LSVT), pharyngeal exercise, gargling, vocal exercises, velar elevation and airway closure/breath holding. The "motor with swallow" category includes Mendelssohn, super supraglottic, supraglottic, effortful, tongue holding and swallow (frequency). The "Sensorial" category includes bolus effects (volume, viscosity, temperature, taste enhancement) and stimulation (thermal-tactile stimulation, electrical stimulation, occluding trach and visual feedback).

This article aims to describe a swallowing management and investigated associated factors with swallowing functionality in a case series of patients with Parkinson's Disease (PD).

Material and Methods

Selection of patients

This is a prospective and retrospective long-term study of 24 dysphagic patients with idiopathic PD, observed in a five year period (2006-2011) in a large Brazilian university hospital. It was an open case series and during the five years of study there were patients moving in and out.

Only the patients who had at least three evaluations, complaints of swallowing and regular check-ups with Outpatient Neurology were included in the study. Patients with concomitant diseases or disorders able to cause dysphagia were excluded from the analysis.

Procedures

The patients underwent swallowing evaluation, Functional Oral Intake Scale (FOIS) and therapeutic intervention every three months.

The swallowing evaluation was obtained through a Fiberoptic Endoscopic Evaluation of Swallowing (FEES) and clinical evaluation. In both evaluations, three types of food were offered: 1) Lemon juice colored with green dye. 2) Nectar, honey and pudding consistencies, all colored with green dye (these fluids were obtained with the addition of 2, 3 and 4 teaspoons of a thickener (Thicken-easy®) to 100 mL of water, respectively, and were offered in two different quantities, 3 mL and 7 mL). 3) A solid consistency was represented by a cornstarch biscuit.

The food was given to patients in the following sequence: liquid and nectar (3 mL, 3 mL, 7 mL and 7 mL); honey (3 mL, 3 mL, 7 mL and 7 mL); pudding (2 tablespoons); solid (½ cornstarch biscuit). The liquid food was administered in 20 mL syringes, with the sample introduced into the patient's oral cavity. As difficulties in swallowing were observed, protective maneuvers of the airways and/or changes in head posture were performed in order to assist oral feeding in a safe way.

FEES's were carried out by an otolaryngologist while the food was offered by a speech-language therapist.

In the clinical evaluation, as the patients swallowed, cervical auscultation was performed to identify abnormal signs at the pharyngeal swallowing phase. Oral bolus

transit time, anterior or posterior escape, positive cervical auscultation (with signs that indicate a presence of stasis or penetration with aspiration risk), coughing (before, during or after swallowing) and wet voice were also observed.

Based on the clinical evaluation and FEES, the FOIS[13] was applied. The FOIS ranks patients into levels. For the present study the levels are described as follows: Level 1) Nothing by mouth. Level 2) Tube dependent with minimal attempts of food or liquid. Level 3) Tube dependent with consistent oral intake of food or liquid. Level 4) Total oral diet of one or two consistencies (nectar and honey, honey and pudding). Level 5a) Total oral diet with multiple consistencies but with restriction of two consistencies (for example, solid and liquid), with or without compensation. Level 5b) Total oral diet with multiple consistencies but with restriction of one consistency (for example, solid or liquid), with or without compensation. Level 5c) Total oral diet with multiple consistencies, but requiring compensation. Level 6) Total oral diet with multiple consistencies without special preparation but with specific food limitations (for example: fibers, grains and some vegetables) and speed and volume modification if necessary. Level 7) Total oral diet with no restriction.

To analyze the swallowing functionality, patients were observed until the point of the time at which a fall or rise in FOIS was noted.

Following these swallowing evaluations, every three months, patients received therapeutic intervention regarding adequate food consistency and volume, besides maneuvers or exercises to improve swallowing functionality. They were oriented to perform the maneuvers daily and received written instructions for each one.

In the “compensatory” category the maneuvers used were: 1) Chin-tuck: to improve airway protection during swallowing. 2) Bolus consistency: to facilitate the feeding of

patients with decreased coordination of tongue, reduced contraction of pharynges, delay in triggering swallowing reflex, reduced airway protection and chewing difficulty.

In the "motor without swallow act" category were used: 1) Strengthening-tongue: to increase strength to eject the bolus. 2) Tongue control: to improve tongue mobility and facilitate the bolus management in the oral cavity. 3) Shaker: to increase strength in supra hyoid muscles reducing the penetration and aspiration risk due to stasis in pyriform sinus. 4) Vocal exercises: to improve airway protection though the improvement in the glottis adduction.

In the "motor with swallow act" category were used: 1) Effortful swallow: to increase strength to eject the bolus and to approximate the larynx structures, improving airway protection. 2) Tongue holding (Masako maneuver): to increase movements of pharyngeal muscles against the basis of the tongue during the act of swallowing. 3) Frequency of swallowing (multiple swallows): to clear stasis.

In the "sensorial" category were used: 1) Bolus effects: changes in volume, viscosity, temperature or taste in order to improve oral and pharyngeal sensibility and control bolus management.

Data Analysis

The patients were classified according to changes of swallowing functionality measured by FOIS. Three groups were observed: improved, stayed the same and worsened.

In order to investigate factors related to swallowing functionality, the Chi-square, Kruskal-Wallis and Fisher tests were used. The gender and age at first evaluation, age at onset of symptoms and disease duration (disease duration = time between the onset of PD

symptoms and the first evaluation) were considered as independent variables. The swallowing functionality over time was described by Kaplan-Meier Survival Analysis.

The statistical analysis was performed using the Statistical Package for the Social Sciences (SPSS) software version 13.0 for Windows and p-values lower than 0.05 were considered significant.

This study was approved by the Ethic Board Committee of the Faculty of Medical Sciences at the University of Campinas, Brazil (Protocol Number 796/2005).

Results

The group of 24 patients was constituted by 16 men and 8 women. The average age of onset of PD symptoms was 53.8 (± 6.5) years old. The average age for the first evaluation was 65.4 (± 8.6) years old. The average disease duration was 139.2 (±65) months, in other words, an average of 11 years between the first symptoms and the first evaluation for swallowing management.

Ten patients improved, five stayed the same and nine worsened their swallowing functionality during follow-up. The characteristics of these groups are described in Table 1. There was no statistically significant difference between the groups.

No significant association with swallowing functionality in this case series was found.

Figure 1 shows the patients whose swallowing worsened. They gradually lost swallowing functionality. According to Kaplan-Meier analysis, the patients had 17% probability of worsened swallowing functionality at the tenth month of follow-up.

Figure 2 shows the patients with improvement. The majority had improved swallowing functionality up to the tenth month. According to the Kaplan-Meier analysis, in

ten months of follow-up the probability of improvement in swallowing functionality was 44%.

The frequencies of maneuvers indicated in the therapeutic intervention are shown in Table 2. The maneuvers chin-tuck, bolus consistency, strengthening–tongue, vocal exercises, swallow frequency and bolus effect were suggested to 50% or more of the patients.

Table 1. Descriptive analysis of clinical aspects of a case series of patients with PD followed in a dysphagia outpatient between 2006 and 2011, stratified by swallowing functionality (N=24).

	Worsened (N=9)		*Stayed the same (N=5)*		*Improved (N=10)*	
Variables	*Min-Max*	*Median*	*Min-Max*	*Median*	*Min-Max*	*Median*
Gender (male/female)	2/7		3/2		4/6	
Age at first evaluation (years)	51-78	64	52-75	66	53-81	61
Age at symptoms' onset (years)	39-74	57	49-65	52	49-75	53
Disease duration (months)	24-204	48	36-204	168	24-144	78
Time up to worsened swallowing (months)[a]	7-78	33	-	-	-	-
Time of follow-up (months)	-	-	15-37	30	-	-
Time up to improved swallowing (months)[b]	-	-	-	-	1-67	10

[a] Time estimated from the onset of PD symptoms.

[b] Time estimated from the first evaluation.

Table 2. Frequency of maneuvers recommended in the therapeutic intervention in a case series of patient with PD followed in a dysphagia outpatient between 2006 and 2011 (N= 24).

Categories	**Maneuvers**	**N (%)**
Compensatory	Chin-tuck	16 (66.7)
	Bolus consistency	19 (79.2)
Motor without swallow act	Strengthening–tongue	16 (66.7)
	Tongue control	10 (41.7)
	Shaker	3 (12.5)
	Vocal exercises	12 (50.0)
Motor with swallow act	Effortful	10 (41.7)
	Tongue holding	4 (16.7)
	Swallow Frequency (Multiple swallows)	19 (79.2)
Sensorial	Bolus effects	20 (83.3)

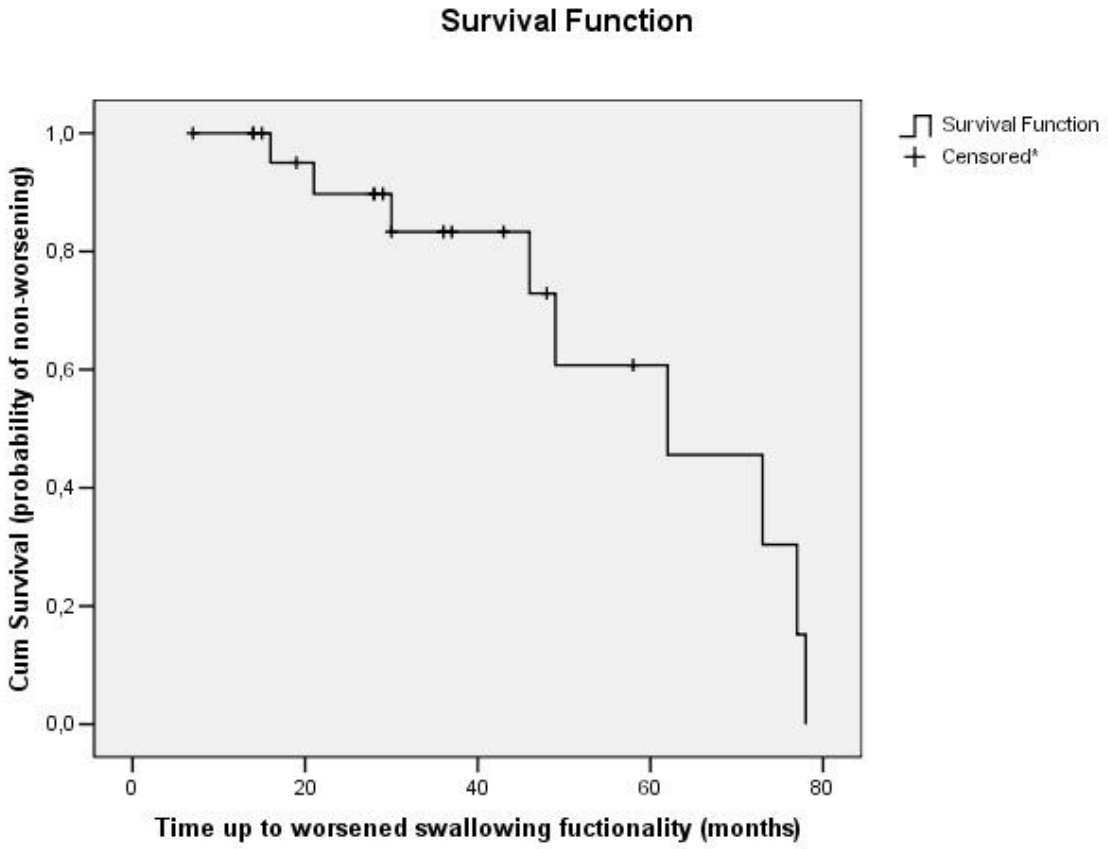


Figure 1. Survival plot considering **worsened swallowing** according to FOIS levels in PD patients followed at a dysphagia outpatient between 2006 and 2011 (N=24). *Censored observation shows patients lost in follow-up or patients without worsened swallowing functionality during the observation period.

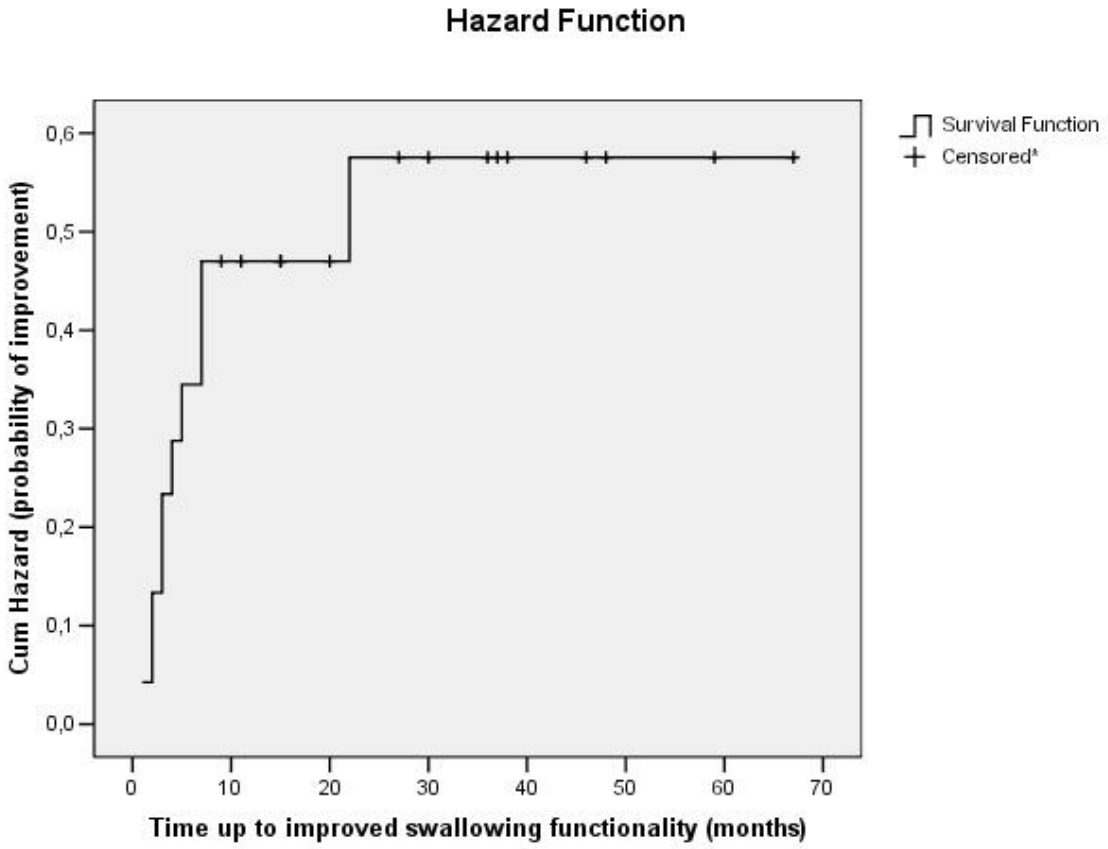


Figure 2. Hazard plot considering the **improved swallowing** according to FOIS levels in PD patients followed at a dysphagia outpatient between 2006 and 2011 (N=24). *Censored observation shows lost patients in follow-up or patients without improved swallowing functionality during the observation period.

Discussion

In this case series an average of 11 years between the PD onset and the patient's first evaluation at the dysphagia outpatient clinic was observed. This could be explained by several factors, including lack of information or latency between the PD onset and the beginning of swallowing complaints. The literature indicates that the delay in seeking for professional assistance may be damaging, especially because dysphagia is prevalent in long-standing PD, but may be subclinical specially in the early course of the disease.[5,14] Objective swallowing evaluations have repeatedly found impaired swallowing in over than 50% of patients with PD who reported no swallowing abnormalities.[14] During the disease course, 75% to 97% of the patients will suffer from dysphagia.[15-17] Because of these evidences it is important to pay attention to swallowing, always looking for weight loss, malnutrition and pulmonary aspects in order to avoid dysphagia complications.

Müller et al.[9], in a postmortem study found a median age at PD onset of 60 years, a median survival time of approximately 14 years and a dysphagia latency of 10 years. The authors showed that disease duration is important because the latency of dysphagia complaints was positively correlated with the total survival time.

The present study found no association between gender and swallowing functionality in PD. It was previously investigated in United States and a relationship was not found between bronchoaspiration and gender.[20]

In this current study maintenance or improvement of swallowing functionality was observed in the majority of the patients during the follow-up period. There was no statistic difference found between the groups that stayed the same, improved or worsened the swallowing functionality. However, we observed in the descriptive analyses that the group

that worsened had the onset of PD symptoms at a later age, having shorter disease duration at the time of the first evaluation.

These results were observed by other authors. Lorefalt et al.[21] reported a significant reduction of solid food in older patients with PD. The non-ingestion of the solid food per se indicate a lost in FOIS. In PD, higher mortality is associated with some aspects such as: older age, dysphagia and late diagnosis.[22] The older onset is associated with a negative impact on patient's survival.[23]

The group that improved the dysphagia status had its first evaluation younger than the other groups. The early swallowing management may positively affect swallowing functionality. According to the literature, the compensatory maneuvers used in swallowing management can improve airway protection and reduce dysphagia complications.[24] Manor et al.[8] found that in PD the swallowing management in earlier stages of dysphagia was able to prevent aspiration pneumonia and help in the maintenance the quality of life.

Despite of a lack of blind controlled studies and besides the PD being progressive and neurodegenerative, there are indications that improvement can be brought by relatively simple interventions.[25,26] The patients in the studied group showed improvement in the first ten months. According to Robbins[12], some maneuvers need to be performed repeatedly to promote neuroplasticity and improve the swallowing functionality. For the present study, patients received a therapeutic intervention at three-month intervals. If the study had had smaller intervals of observation, faster improvement might have been obtained.

Swallowing Management in Parkinson's disease

This swallowing management aimed to improve the swallowing act as much as possible and compensate for what cannot be solved, with a view to save deglutition.

As a result of this study, it was found that one of the most recommended interventions in PD is compensatory. This category of maneuvers is designed to redirect the bolus away from the airway, without changing airway physiology. According to the literature, thickening liquids to a nectar or honey consistency was used because it exhibited an immediate effect.[28] Chin-tuck maneuver was frequently recommended for swallowing thin liquids.[29]

In PD, there are evidences in literature showing that the chin-tuck maneuver combined with thick liquid can be important in preventing pneumonia.[20]

The bolus consistency was indicated when others maneuvers were inefficient to maintain oral feeding and it was necessary to avoid the consistencies that were hazardous for the patient.

For all the patient of this study, the bolus effect maneuver was indicated when the lack of oral control made it difficult to swallow quickly and in greater volumes. The bolus consistency, bolus effect, changes in the consistency, taste or temperature of the bolus improve oral intake and prevent aspiration.

In the maneuver category "motor without swallow act", the exercises of strengthening tongue and tongue control were frequently indicated because tongue movements of PD patients, especially those responsible for propulsion and chewing, are

considered hypokinetic in the oral phase. In these patients the swallowing oral phase is usually longer and even slower in the pharyngeal phase.[30]

There are evidences that vocal exercises improve the cough function and tongue mobility during swallowing.[27]

Frequency or multiple swallowing, in the maneuver category "motor with swallow act" was performed during the daily feeding, with the aiming to clear stasis and reduce the chance of larynx penetration or aspiration.

Conclusion

The swallowing functionality in this case series was characterized by maintenance or improvement, especially in the first tenth months of follow-up. There was no associated factor with swallowing functionality.

The swallowing management was featured by compensatory maneuvers, motor exercises with and without the act of swallowing.

The improvement found in the present study suggests that swallowing management might positively affect the swallowing functionality of PD patients. It can possibly contribute to a longer period of oral feeding with safe swallowing.

References

1. Groher ME, Crary MA. Dysphagia: clinical management in adults and children. St Louis: Mosby, 2009.

2. Schrag A, Ben-Shlomo Y, Quinn NP. Cross sectional prevalence survey of idiopathic Parkinson's disease and Parkinsonism in London. BMJ 2000; 321:21–2.

3. Rijk MC, Breteler MM, Graveland GA, Ott A, Grobbee DE, Van Der Meche´ FG, et al. Prevalence of Parkinson's disease in the elderly: the Rotterdam study. Neurology 1995; 45:2143–6

4. Jankovic J. Pathophysiology and assessment of parkinsonian symptoms and signs. In: Pahwa R, Lyons K, Koller WC, eds. Handbook of Parkinson's disease. New York: Taylor and Francis Group LLC, 2007. p 79–104.

5. Potulska A, Friedman A, Krolicki L, Spychala A. Swallowing disorders in Parkinson's disease. Parkinsonism Relat Disord 2003; 9:349–53.

6. Dawes C, Kubienciec K. The effects of prolonged gum chewing on salivary flow rate and composition. Arch Oral Biol 2004; 49:665–9.

7. Marik PE, Kaplan D. Aspiration pneumonia and dysphagia in the elderly. Chest. 2003; 124:328-36.

8. Manor Y, Giladi N, Cohen A, Fliss DM, Cohen JT. Validation of a swallowing disturbance questionnaire for detecting dysphagia in patients with Parkinson's disease. Mov Disord 2007; 22:1917–21.

9. Müller J, Wenning GK, Verny M, McKee A, Chaudhuri KR, Jellinger K, et al. Progression of Dysarthia and dysphagia in postmortem-confirmed Parkisonian Disorders. Arch Neurol 2001; 58:259-64.

10. D'Amelio M, Ragonese P, Morgante L, Reggio A, Callari G, Salemi G, et al. Long-term survival of Parkinson's disease: a population-based study. J Neurol 2006;253:33–37.

11. Schüpbach MW, Welter ML, Bonnet AM, Elbaz A, Grossardt BR, Mesnage V, et al. Mortality in patients with Parkinson's disease treated by stimulation of the subthalamic nucleus. Mov Disord 2007; 22:257–61.

12. Robbins J, Butler SG, Daniels SK, Gross RD, Langmore S, Lazarus CL, et al. Swallowing and dysphagia rehabilitation: translating principles of neural plasticity into clinically oriented evidence. Journal of Speech, Language, and Hearing Research 2008 February, S276-S300.

13. Crary MA, Mann GDC, Groher ME. Initial pychometric assessment of a functional oral intake scale for dysphagia in stroke patients. Arch Phys Med Rehabil 2005; 86:1516-20.

14. Miller N, Noble E, Jones D, Burn D. Hard to swallow: dysphagia in Parkinson's disease. Age Ageing 2006; 35:614-18.

15. Logemann JA, Blonsky ER, Boshes B. Editorial: dysphagia in parkinsonism. JAMA 1975; 231:69-70.

16. Stroudley J, Walsh M. Radiological assessment of dysphagia in Parkinson's disease. Br J Radiol 1991; 64:890-3.

17. Bird MR, Woodward MC, Gibson EM, Phyland DJ, Fonda D. Asymptomatic swallowing disorders in elderly patients with Parkinson's disease: a description of findings on clinical examination and videofluoroscopy in sixteen patients. Age Ageing 1994; 23(3):251–54.

18. Miller N, Allcock L, Hildreth AJ, Jones D, Noble E, Burn DJ. Swallowing problems in Parkinson disease: frequency and clinical correlates. J Neurol Neurosurg Pychiatry 2009; 80:1047-9.

19. Baldeschi M, Di Carlo A, Rocca WA; for ILSA Working Group. Italian longitudinal study on aging Parkinson's disease and parkinsonism in a longitudinal study: two-fold higher incidence in men. Neurology 2000; 55:1358-63.

20. Logmann JA, Gensler G, Robbins J, Lindblad AS, Hind JA, Kosek S, et al. A randomized study of three interventions for aspiration of thin liquids in patients with dementia or Parkinson's disease. J Speech Lang Hear Res 2008; 51;173-83.

21. Lorefält B, Gránerus AK, Unosson M. Avoidance of solid food in weight losing older patients with Parkinson's disease. J Clin Nurs 2006; 15(11):1404-12.

22. Lo RY, Tanner CM, Albers KB, Leimpeter AD, Fross RD, Bernstein AL, et al. Clinical features in early Parkinson disease and survival. Arch Neurol 2009; 66(11):1353-8

23. Auyeung M, Tsoi TH, Mok V, Cheung CM, Lee CN, Li R, et al. Ten year survival and outcomes in a prospective cohort of new onset Chinese Parkinson's disease patients. J Neurol Neurosurg Psychiatry 2012; 83(6):607-11.

24. Robbins J, Gensler G, Hind J, Logemann JA, Lindblad AS, Brand D, et al. Comparison of 2 interventions for liquid aspiration on pneumonia incidence a randomized trial. Ann Intern Med 2008; 148(7):509–18.

25. El Sharkawi A, Ramig L, Logemann JA, B.R. Pauloski, A.W. Rademaker, C.H. Smith, et al. Swallowing and voice effects of Lee Silverman Voice Treatment (LSVT(R)): a pilot study. J Neurol Neurosurg Psychiatry 2002; 72:31–6.

26. Hockstein NG, Samadi DS, Gendron K, Handler SD. Sialorrhea: a management challenge. Am Fam Physician 2004; 69: 2628–34.

27. Russell JA, Ciucci MR, Connor NP, Schallert NT. Targeted exercises therapy for voice and swallow in persons with Parkinson's Disease. Brain Res 2010; 1341:3-11.

28. Logemann, J.A. Evaluation and treatment of swallowing disorders (2nd ed.). Austin, TX: Pro-Ed, 1998.

29. Chiappetta ALM, Oda AL. Doenças neuromusculares. In: Ferreira LP, Befi-Lopes DM, Limongi SCO. Tratado de Fonoaudiologia. São Paulo: Roca, 2004, 330-41.

30. Menezes C, Melo A. Does levodopa improve swallowing dysfunction in Parkinson's disease patients? J Clin Pharm Therapeut 2009 34:673–676.

CONCLUSÕES E REFERÊNCIAS BIBLIOGRÁFICAS

CONCLUSÕES

Geralmente, a dor ao engolir não é um aspecto muito enfatizado na avaliação e no tratamento em deglutição de pacientes com ELA. Entretanto, este estudo revela que deve ser dada especial atenção a esta queixa, pois pode indicar uma piora na disfagia capaz de trazer complicações que podem levar a morte.

Como fatores que influenciam na progressão da disfagia na ELA, observou-se que os pacientes com ELA de início bulbar e os pacientes com início tardio dos sintomas tiveram uma progressão mais rápida da disfagia.

Também na ELA, o sexo feminino, a idade de início avançada e o curto período entre o início dos sintomas da doença e a primeira avaliação foram identificados como fatores que levaram à necessidade de indicação de via alternativa de alimentação em menor tempo.

Não foram identificados fatores que influenciassem a progressão da disfagia na DP, no entanto, observou-se melhora ou estabilização da deglutição na maioria dos sujeitos estudados, principalmente nos dez primeiros meses de acompanhamento.

Quanto à intervenção fonoaudiológica na DP foram mais indicados os exercícios para força de ejeção e mobilidade de língua, deglutições múltiplas, exercícios vocais e manobras compensatórias.

REFERÊNCIAS BIBLIOGRÁFICAS

1. Groher ME, Crary M. Dysphagia: Clinical management in adults and children. MO: Mosby, 2009.
2. Schrag A, Ben-Shlomo Y, Quinn NP. Cross sectional prevalence survey of idiopathic Parkinson's Disease and Parkinsonism in London. BMJ 2000;321:21–2.
3. Rijk MC, Breteler MM, Graveland GA, Ott A, Grobbee DE, Van Der Meche´ FG, et al. Prevalence of Parkinson's disease in the elderly: the Rotterdam study. Neurology 1995; 45:2143–6
4. O'toole O, Traynor BJ, Brennan P et al. Epidemiology and clinical features of amyotrophic lateral sclerosis in Ireland between 1995 and 2004. J. Neurol Neurosurg Psychiatry 2008; 79:30-2.
5. Worms PM. The epidemiology of motor neuron diseases: a review of recent studies. J Neurol Sci 2001, 191:3-9.
6. Fattori B, Grosso M, Bongioanni P, et al. Assessment of swallowing by oropharyngoesophageal scintilography in patients with amyotrophic lateral sclerosis. Dysphagia 2006; 21:280-6.
7. Hadjikoutis S, Wiles CM. Respiratory complications related to bulbar dysfunction in motor neuron disease. Acta Neurol Scand 2001; 103:207-13.
8. Oliveira ASB, Pereira RDB. Amyotrophic Lateral Sclerosis (ALS): three letters that change the people's life. Arq Neuropsiquiatr 2009; 67(3-A):750-82.
9. Ganzini L, Johnston WS, Hoffman WF Correlates of suffering in amyotrophic lateral sclerosis. Neurology 1999; 52:1434–40.

10. Schrag A, Ben-Shlomo Y, Quinn NP. Cross sectional prevalence survey of idiopathic Parkinson's disease and Parkinsonism in London. BMJ 2000; 321:21–2.

11. Jankovic J. Pathophysiology and assessment of parkinsonian symptoms and signs. In: Pahwa R, Lyons K, Koller WC, eds. Handbook of Parkinson's disease. New York: Taylor and Francis Group LLC, 2007. p 79–104.

12. Potulska A, Friedman A, Krolicki L, Spychala A. Swallowing disorders in Parkinson's disease. Parkinsonism Relat Disord 2003; 9:349–53.

13. Dawes C, Kubienciec K. The effects of prolonged gum chewing on salivary flow rate and composition. Arch Oral Biol 2004; 49:665–9.

14. Müller J, Wenning GK, Verny M, McKee A, Chaudhuri KR, Jellinger K, et al. Progression of Dysarthia and dysphagia in postmortem-confirmed Parkisonian Disorders. Arch Neurol 2001; 58:259-64.

15. Marik PE, Kaplan D. Aspiration pneumonia and dysphagia in the elderly. Chest. 2003; 124:328-36.

16. Manor Y, Giladi N, Cohen A, Fliss DM, Cohen JT. Validation of a swallowing disturbance questionnaire for detecting dysphagia in patients with Parkinson's disease. Mov Disord 2007; 22:1917–21.

17. Logemann J, Blonsky ER, Boshes B. Lingual control in Parkinson's disease. Trans Am Neurol Assoc 1973;98:276–278.

18. Ertekin C, Aydogdu I, Yucear N. Piecemeal deglutition and dysphagia limit in normal subjects and in patients with swallowing disorders. J Neurol Neurosurg Psychiatry 1996;61:491–6.

19. Ertekin C, Aydogdu I, Yuceyar N, Tarlaci S, Kiylioglu N, Pehlivan M, Celebi G. Electrodiagnostic methods for neurogenic dysphagia. Electroenceph Clin Neurophys 1998;109:331–40.

20. Ertekin C, Palmer J. Physiology and electromygraphy of swallowing disorders. Clin Neurophys 2000;53(Suppl):148–60.

21. Ertekin C, Tarlaci S, Aydogdu I, Kiylioglu N, Yuceyar N, Turman AB, et al. Electrophysiological evaluation of pharyngeal phase of swallowing in patients with Parkinson's disease. Mov Disord 2002;17:942–9.

22. Leder SB, Novella S, Patwa H. Use of Fiberoptic Endoscopic Evaluation of swallowing (FEES) in patients with Amyotrophic Lateral Sclerosis. Dysphagia 2004; 19:177–81.

23. Ludolph AC, Prosiegel M, Riecker A. Diagnosis and treatment of bulbar symptoms in amyotrophic lateral sclerosis. Nat Clin Pract Neurol 2008; 4(7):366-74.

24. Kawai S, Tsukuda M, Mochimatsu I, Enomoto H, Kagesato Y, Hirose H et al. A study of the early stage of dysphagia in amyotrophic lateral sclerosis. Dysphagia 2003; 18:1–8.

25. Solazzo A, Vecchio LD, Reginelli A, Monaco L, Sagnelli A, Monsorrò M et al. Search for compensation postures with videofluoromanometric investigation in dysphagic patients affected by Amyotrophic Lateral Sclerosis. Radiol med 2011, 116:1083–94.

26. Higo R, Tayama N, Nito T. Longitudinal analysis of progression of dysphagia in Amyotrophic Lateral Sclerosis. Auris Nasus Larynx 2004; 31:247-54.

27. Sordi MD, Mourão LF, Da Silva AA, Flosi LCL. Importância da interdisciplinaridade na avaliação das disfagias: avaliação clínica e videofluoroscópica da deglutição. Rev Bras Otorrinol 2009; 75(6):776-87.

ANEXOS

ANEXO I – PROCOLO DE AVALIAÇÃO DA DEGLUTIÇÃO

Protocolo de Avaliação Clinica e Videoendoscópica da Deglutição (VED)

(Nov/11)

Data da Avaliação:____/____/_____
Paciente:_____
Registro:_____DN:_____Idade:_____
Endereço:_____
Tel:_____Informante:_____

Aspectos da Linguagem: ( ) alteração na produção da fala ( ) alteração na organização da linguagem ( ) CSA
Habilidades Linguisticas alteradas: ( ) pragmático-discursivo ( ) fonético-fonológico ( ) sintático-semântico

H.D.:_____
HPQ:_____
_____
_____
Primeiros sintomas da doença:_____
Data de inicio dos primeiros sintomas:_____
Tempo de Doença:_____
Condições Gerais do Paciente: _____
_____
Medicamentos:_____
_____
Queixa atual:_____
_____
_____
Queixa de Deglutição: ( )Sim ( )Não
Tempo de Queixa da deglutição_____
Hábitos Alimentares Pregressos:_____
_____
_____
_____
Recordatório Alimentar (24 hrs):_____
_____
_____
_____
_____
_____Peso Usual:_____
Peso Atual: _____ Altura:_____ IMC: Peso/altura^2: _____
Renda mensal Familiar: _____ Nº integrantes familia: _____

Estado Geral de Saúde: ( ) Sim ( ) Não
Cardiopatias: ( ) não ( ) sim Especificar: _____
Pressão alta: ( ) não ( ) sim Especificar: _____
Infecções pulmonares: ( ) não ( ) sim Especificar: _____
Alterações gástricas: ( ) não ( ) sim Especificar: _____
Alterações bucais e dentição: ( ) não ( ) sim Especificar: _____
Desnutrição: ( ) não ( ) sim
Desidratação: ( ) não ( ) sim
Diabetes: ( ) não ( ) sim Especificar tipo: _____

Traqueostomia: ( ) Presente ( ) ausente
Cânula: ( ) metal ( ) plástico PVC ( ) silicone ( ) c/ Cuff ( ) s/ cuff
SNE: ( ) Presente ( ) ausente

Queixas de deglutição:

FASES DA DEGLUTIÇÃO							
FASE ANTECIPATÓRIA							
Prazer em se alimentar	S N						
Apetite	S N						
FASE ORAL		**Freq.**	**Duração**	**FASE FARÍNGEA**		**Freq.**	**Duração**
Alimento escapa da boca	S N			Refluxo nasal	S N		
Dificuldade para mastigar	S N			Tosse	Antes S N Durante S N Após S N		
Alimento gruda no céu da boca	S N			Engasgos	Antes S N Durante S N Após S N		
Dificuldade para empurrar o alimento	S N			Pigarro	Antes S N Durante S N Após S N		
Restos de alimentos na CO	S N			Sensação de alimento parado	S N		
Líquido / saliva escapa da boca	S N			Dificuldade para engolir	S N		
Dor na cavidade oral	S N			Dor para engolir	S N		
Dificuldade para deglutir saliva	S N			Demora para engolir	S N		
				Dor no peito	S N		

Avaliação clínica indireta

Face	**Mandíbula**
Mobilidade (VII): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Sensibilidade (V): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red	Mobilidade (V): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Sensibilidade (V): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Reflexo de mordida (V): ( ) pres ( ) aus
Lábios	**Língua**
Mobilidade (VII): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Sensibilidade (V): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red	Mobilidade (XII): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Sensibilidade (V,IX): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Gustação (VII, IX): ( )pres ( ) aus ( ) red
Orofaringe	**Laringe**
Mobilidade (X): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Voz hipersanal(X): ( ) pres ( ) aus Sensibilidade (IX, V): D ( ) pres ( ) aus ( ) red E ( ) pres ( ) aus ( ) red Reflexo de vômito - língua (IX): D ( ) pres ( ) aus E ( ) pres ( )aus - palato (V): D ( ) pres ( )aus E ( ) pres ( ) aus Prova de Gutzman: ( ) pres ( ) aus	Mobilidade (X): ( ) nl ( ) alt ( ) red Voz /a/: G R B A S I – Voz (Fala): G R B A S I Voz Molhada: ( ) pres ( ) aus Elevação laríngea (X, IX): ( )pres ( ) aus ( ) red Reflexo de tosse (X): ( ) pres ( ) aus Tosse (X): ( ) Eficiente ( ) Ineficiente
Ausculta Laríngea	
( ) nl ( ) alt	

Avaliação clínica direta

	Consistência																	
	Líquido					Néctar					Mel					Pastoso		Sólido
	3	3	7	7	Liq Cont	3	3	7	7	Liq Cont	3	3	7	7	Liq Cont	sob	sob	
Mastigação																		E IN
TOL																		
Número de deglutições																		
Escape Anterior																		
Escape Posterior																		
Restos de alimentos																		
Refluxo nasal																		
Tosse voluntária																		
Tosse reflexa																		
Tosse antes																		
Tosse durante																		
Tosse após																		
Voz molhada																		
Redução da elevação laringea																		
Ausculta laringea																		

Legenda: P = presente; A = ausente; nl = normal ; alt = alterado

Sinais de penetração/aspiração:
Alteração coloração facial: _____
Alteração da freqüência respiratória: _____
Alteração da saturação O_2: _____
Manobras Utilizadas: _____

Classificação Avaliação Clínica

	Silva (1999)		Estágio	Yorkston et al (2004) – ELA
	Deglutição Normal	Ausência de alteração nas fases oral e faríngea da deglutição	1	Sem Alteração
	Disfagia Leve	Controle e transporte do bolo alimentar atrasado e lento, sem sinais de penetração laríngea na ausculta	2	Tempo de refeição aumentado, fadiga para mastigar, dificuldades no transporte do bolo, Presença de espasticidade de língua e flacidez faríngea. Redução da ingesta de líquido.
	Disfagia Moderada	Controle e transporte do bolo alimentar atrasado e lento, com sinais de penetração laríngea e risco de aspiração	3	Mudança de consistência alimentar. Adição de molhos ou pela trituração e homogeinização dos alimentos.
	Disfagia Severa	Presença substancial e ausência ou falha completa do bolo alimentar	4	Indicação de sonda de alimentação com complementação de alimentos via oral.
			5	Suspensão completa da alimentação oral. Via alternativa exclusiva e controle das secreções

Avaliação Videoendoscópica da Deglutição

1.Fossas Nasais
Septo: Centrado( ) desvio D ( ) desvio E ( ) irregularidades não obstrutivas ( )
Mucosa: Pálida ( ) edemaciada ( ) úmida ( ) atrófica ( )
Cornetos: normotróficos ( ) hipertróficos ( )

2.Rinofaringe
Mucosa: Pálida ( ) edemaciada ( ) úmida ( ) atrófica ( )
Óstios tubários: livres ( ) obstruidos ( )

3.Esfíncter Velo-Faríngeo
Fonação:
fechamento completo ( ) fechamento incompleto ( ) coronal ( ) sagital ( ) circular ( ) circular-Passavant ( )
insuficiente ( ) incompetente ( )
Deglutição:
fechamento completo ( ) fechamento incompleto ( ) coronal ( ) sagital ( ) circular ( ) circular-Passavant ( )
insuficiente ( ) incompetente ( )

4.Hipofaringe (IX, X, XII)
Base de lingua mobilidade: adequada ( ) alterada ( )_____
Parede posterior mobilidade: adequada ( ) alterada ( )_____
Valécula: normal ( ) lesão ( ) estase salivar ( )
Epiglote: normal ( ) ômega ( ) lesão ( )
Aritenóides: normal ( ) hiperemia ( ) edema ( )
Região inter-aritenóidea: normal ( ) hiperemia ( ) edema ( )
Recessos Piriformes: livres ( ) obliterados ( ) estase salivar D ( ) E ( )
Sensibilidade: normal ( ) diminuida ( ) ausente ( )
Mucosa: normal ( ) edemaciada ( ) rugosidade ( ) paquidermia ( )

5.Laringe
Pregas Vocais
Móveis ( )
Paresia D ( ) E ( )
Imobilidade ( )
Arqueamento ( )
Atrofia ( )
Lesão_____ D ( ) E ( )
Outros_____

Pregas Vestibulares
Normais ( )
Hiperconstrição D ( ) E ( )
Assimetria Laringea Sim ( ) Não ( )

Sensibilidade Laringea ao estímulo mecânico:
Epiglote: normal ( ) alterado ( )
Pregas Vocais: normal ( ) alterado ( )
Subglote: normal ( ) alterado ( )
Prega ariepglótica: normal ( ) alterado ( )
Aspiração de Saliva: presente ( ) ausente ( )

6.Fechamento Glótico
Completo ( ) Incompleto ( )
Fenda triangular posterior ( )
Fenda triangular médio-posterior ( )
Fenda fusiforme anterior ( )
Fenda fusiforme toda extensão ( )
Fenda em ampulheta ( )

Avaliação Videoendoscópica da Deglutição (VED)

CONSISTÊNCIA		Líquido					Liquido Engrossado										Pastoso		Sólido	
		Líquido					Néctar					Mel					Pudim		Sólido	
SEQUENCIA																				
QUANTIDADE		3ml	3ml	7ml	7ml	Contínuo ( )copo ( )canudo	3ml	3ml	7ml	7ml	Contínuo ( )copo ( )canudo	3ml	3ml	7ml	7ml	Contínuo ( )copo ( )canudo	Col. Sobr.	Col. Sobr.	Bolacha	Bolacha
Refluxo Nasal																				
Escape	Anterior																			
Dg faríngea	Iniciada																			
Duração T.O.																				
Elevação Laríngea																				
Penetração	Alta	S	S	S	S	S N	S	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N
	V.	S	S	S	S	S N	S	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N
Aspiração																				
Reflexo Tosse		S	S	S	S	S N	S	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N
	Eficiente																			
	N.																			
Resíduos	Dorso																			
	Valécula																			
	S. P. D.																			
	S. P. E.																			
	P. P.																			
	E. E. S.																			
N° Degl.																				
Nº Degl.Clarear																				
Clareamento		S	S	S	S	S N	S	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N	S N
Alim. Orofar.																			T NT	T NT
Manobra																				
Efetividade M																				
Sensibilidade Laríngea	Estrutura estimulada: _____ Presente ( ) Ausente ( ) Reduzida ( ) Estrutura estimulada: _____ Presente ( ) Ausente ( ) Reduzida ( )																			

Legenda: S = Sim; N = Não; Ad = Adequado; Au = Aumentado; D = Diminuído; P = Presente; As = Ausente; R = Reduzido; S.P.D. = Seio piriforme Direito; S.P.E. = Seio Piriforme Esquerdo; P.P. = Parede Posterior; E.E.S. = Esfíncter Esofágico Superior; T = Triturado; NT = Não Triturado; CPB = Cabeça para baixo; CPD = Cabeça para direita; CPE = Cabeça para esquerda; DM = Deglutição Múltipla; DE = Deglutição com Esforço; SG = Supraglótica; SSG = Super Supraglótica; CID: Cabeça inclinada para direita; CIE: Cabeça inclinada para esquerda; M: Mendelsohn; Estase : 0 = Sem estase; C = Aderido à mucosa; 1= Leve; 2= Moderado; 3= Severa

CLASSIFICAÇÃO DA AVALIAÇÃO VIDEOENDOSCÓPICA VED (MACEDO FILHO et al., 2000)		
	Deglutição Normal (Grau 0)	Contenção oral normal, reflexos presentes, ausência de estase salivar, alimentar e aspiração, menos de três tentativas de propulsão para clareamento do bolo;
	Disfagia Leve (Grau 1)	Estase pós-deglutição pequena, menos de três tentativas de propulsão para clareamento do bolo, ausência de regurgitação nasal e penetração laríngea;
	Disfagia Moderada (Grau 2)	Estase salivar moderada, maior estase pós-deglutição, mais de três tentativas de propulsão do bolo, regurgitação nasal, redução da sensibilidade laríngea com penetração, porém sem aspiração laringo-traqueal;
	Disfagia Grave (Grau 3)	Grande estase salivar, piora acentuada de resíduos pós-deglutição, propulsão débil ou ausente, regurgitação nasal, aspiração traqueal.

Functional Oral Intake Scale – FOIS (Crary, MA et al., 2005) - adaptada		
	1	Sem alimentação via oral
	2	Via alternativa de alimentação com ingestão oral mínima para treino
	3	Dieta mista (via oral + via alternativa de alimentação)
	4	Dieta via oral com uma ou duas consistências (néctar e mel, mel e pudim)
	5a	Dieta via oral com compensações, mas sem restrição de consistência
	5b	Dieta via oral com restrição p/ 1 única consistência (ex. líquido ou sólido e/ou necessidade de preparo - ex. amolecido/amassado) com ou sem compensações
	5c	Dieta via oral com restrição para 2 consistências (ex. líquido e sólido) com ou sem compensação
	6	Dieta via oral sem modificação da preparação, com modificação de volume e/ou velocidade, e/ou limitação para alimentos específicos (Ex. grãos, fibra, folhas)
	7	Dieta via oral sem restrição

Conclusão:_____
_____
_____
_____
_____

Conduta:
- Consistências_____
- Manobras_____
- Exercícios / Frequência_____
_____
_____
_____
_____
_____
_____
_____
_____
_____

Avaliador: _____

ANEXO II - FICHA DE RETORNO AO AMBULATÓRIO DE DISFAGIA

FICHA DE EVOLUÇÃO DO PACIENTE — Ficha Nº

NOME:_____ HC: _____

DATA DESTE RETORNO: ___/___/____

Queixa atual: ( ) deglutição. Especifique:_____
( ) fala. Especifique:_____
( ) linguagem. Especifique:_____

Ocorrência: ( ) pneumonia. Especifique (internação/causa/data): _____
( ) internação. Especifique (causa/período):_____
( ) outro. Especifique:_____

Peso atual: _____ Recordatório Alimentar (24h): _____

Avaliação da deglutição realizada: ( ) clínica ( ) videoendoscopia ( ) relato

Descrição:_____

FICHA DE EVOLUÇÃO DO PACIENTE — VERSO

Classificação Avaliação Clínica (Silva, 1999): ( ) deglut. normal ( ) disfagia leve ( ) disfagia moderada ( ) disfagia severa
Classificação Avaliação VED (Macedo Filho, 2000): ( ) deglut. normal ( ) disfagia leve ( ) disfagia moderada ( ) disfagia grave
Estágio ELA (Yorston et AL, 2004): ( ) 1 ( ) 2 ( ) 3 ( ) 4 ( ) 5
Escala FOIS (Crary et al, 2005): ( ) 1 ( ) 2 ( ) 3 ( ) 4 ( ) 5 ( ) 6 ( ) 7

Conduta:
- Consistências_____
- Manobras_____
- Exercícios / Frequência_____

Observações: _____

DATA DO PRÓXIMO RETORNO: ____/____/_____.

ANEXO III – APROVAÇÃO DO COMITÊ DE ÉTICA EM PESQUISA

FACULDADE DE CIÊNCIAS MÉDICAS
COMITÊ DE ÉTICA EM PESQUISA

www.fcm.unicamp.br/fcm/pesquisa

CEP, 26/04/11.
(PARECER CEP: N° 796/2005)

PARECER

I - IDENTIFICAÇÃO:

PROJETO: **"ESTUDO DA CORRELAÇÃO DA DIADOCOCINESIA FONOARTICULATÓRIA E DA DISFAGIA OROFARÍNGEA NA DOENÇA DE PARKINSON, DE MACHADO-JOSEPH E ESCLEROSE LATERAL AMIOTRÓFICA".**

PESQUISADOR RESPONSÁVEL: Lucia Figueiredo Mourão

II – PARECER DO CEP.

O Comitê de Ética em Pesquisa da Faculdade de Ciências Médicas da UNICAMP tomou ciência e aprovou a extensão do projeto de pesquisa com uso de dados já coletados e registrados da avaliação fonoaudiológica, com isso o Termo de Consentimento Livre e Esclarecido pode ser dispensado, referente ao protocolo de pesquisa supracitado.

O conteúdo e as conclusões aqui apresentados são de responsabilidade exclusiva do CEP/FCM/UNICAMP e não representam a opinião da Universidade Estadual de Campinas nem a comprometem.

III – DATA DA REUNIÃO.

Homologado na IV Reunião Ordinária do CEP/FCM, em 26 de abril de 2011.

Prof. Dr. Carlos Eduardo Steiner
PRESIDENTE do COMITÊ DE ÉTICA EM PESQUISA
FCM / UNICAMP

Comitê de Ética em Pesquisa - UNICAMP
Rua: Tessália Vieira de Camargo, 126
Caixa Postal 6111
13083-887 Campinas – SP

FONE (019) 3521-8936
FAX (019) 3521-7187
cep@fcm.unicamp.br

CEP, 27/09/11.
(**PARECER CEP:** N° 796/2005)

PARECER

I - IDENTIFICAÇÃO:

PROJETO: **"ESTUDO DA CORRELAÇÃO DA DIADOCOCINESIA FONOARTICULATÓRIA E DA DISFAGIA OROFARÍNGEA NA DOENÇA DE PARKINSON, DE MACHADO-JOSEPH E ESCLEROSE LATERAL AMIOTRÓFICA".**

PESQUISADOR RESPONSÁVEL: Lucia Figueiredo Mourão

II – PARECER DO CEP.

O Comitê de Ética em Pesquisa da Faculdade de Ciências Médicas da UNICAMP tomou ciência e aprovou a dispensa do Termo de Consentimento Livre e Esclarecido para utilização dos dados já coletados e registrados de todos os sujeitos com Doença de Parkinson e Esclerose Lateral Amiotrófica que já foram atendidos no Ambulatório de Otorrinolaringologia/Disfagia do HC/UNICAMP no período de fevereiro/2006 a dezembro/2011, referente ao protocolo de pesquisa supracitado.

O conteúdo e as conclusões aqui apresentados são de responsabilidade exclusiva do CEP/FCM/UNICAMP e não representam a opinião da Universidade Estadual de Campinas nem a comprometem.

III – DATA DA REUNIÃO.

Homologado na IX Reunião Ordinária do CEP/FCM, em 27 de setembro de 2011.

Prof. Dr. Carlos Eduardo Steiner
PRESIDENTE do COMITÊ DE ÉTICA EM PESQUISA
FCM / UNICAMP

Comitê de Ética em Pesquisa - UNICAMP
Rua: Tessália Vieira de Camargo, 126
Caixa Postal 6111
13083-887 Campinas – SP

FONE (019) 3521-8936
FAX (019) 3521-7187
cep@fcm.unicamp.br

ANEXO IV – DECLARAÇÃO DOS AUTORES

DECLARAÇÃO

Nós, Karen Fontes Luchesi (RG 23004889-9) e Satoshi Kitamura (RG 3087798), declaramos para os devidos fins que as cópias dos artigos, submetidos para publicação em revistas científicas ou anais de congressos sujeitos a arbitragem, que constam nesta tese de doutorado, intitulada "A disfagia na Esclerose Lateral Amiotrófica e na Doença de Parkinson", não infringem os dispositivos da Lei nº 9.610/98, nem o direito autoral de qualquer editora.

Campinas, 06 de fevereiro de 2013.

Karen Fontes Luchesi

Satoshi Kitamura