Auris Nasus Larynx xxx (2016) xxx–xxx

Contents lists available at ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

Dysphagia in Amyotrophic Lateral Sclerosis: Relationships between disease progression and Fiberoptic Endoscopic Evaluation of Swallowing

Bruno Fattori [a,*], Gabriele Siciliano [b], Valentina Mancini [a], Luca Bastiani [c], Paolo Bongioanni [d], Elena Caldarazzo Ienco [b], Maria R. Barillari [e], Salvatore O. Romeo [a], Andrea Nacci [a]

[a] *ENT, Audiology and Phoniatrics Unit, Department of Clinical and Experimental Medicine, University of Pisa, Pisa, Italy*
[b] *Neurological Clinic, Department of Clinical and Experimental Medicine, University of Pisa, Pisa, Italy*
[c] *Institute of Clinical Physiology of the Italian National Research Council (IFC-CNR), Pisa, Italy*
[d] *Neurorehabilitation Unit, Department of Neurosciences, University of Pisa, Pisa, Italy*
[e] *Audiology and Phoniatrics Unit, University of Napoli 2, Napoli, Italy*

ARTICLE INFO

Article history:
Received 17 April 2016
Accepted 5 July 2016
Available online xxx

Keywords:
Dysphagia
Amyotrophic Lateral Sclerosis
Fiberoptic Endoscopic Evaluation of Swallowing
ALSFRS
b-ALSFRS

ABSTRACT

Objective: Our aim was to evaluate the relationship between the disease severity of Amyotrophic Lateral Sclerosis (ALS) and the following parameters of Fiberoptic Endoscopic Evaluation of Swallowing (FEES): premature spillage, post-swallowing residue and aspiration.
Methods: We studied 202 patients (95 women and 107 men) with ALS; of these, 136 had spinal and 66 had bulbar onset. They were analyzed according to the Amyotrophic Lateral Sclerosis Functioning Rating Scale (ALSFRS) and the b-ALSFRS subscale (bulbar scale). All subjects underwent FEES. Post-swallowing residue was classified into four classes (0–3); premature spillage and aspiration were considered either present or absent.
Results: Spearman's correlation test showed a highly significant correlation ($p < 0.0001$) between the value of ALSFRS and b-ALSFRS and the FEES parameters as the following: disease severity and dysphagia severity are closely related, both in spinal and bulbar onset, no matter what bolus texture was used. Spearman's Rho was more significant for post-swallowing residue, ≤ -0.500 with all three consistencies ($p < 0.0001$) in spinal onset and -0.520 only with liquid bolus ($p < 0.0001$) in bulbar onset. Independent *T*-Test revealed a significant difference ($p < 0.0001$) between the mean ALSFRS and b-ALSFRS scores and the presence/absence of aspiration. For the premature spillage in spinal onset (ALSFRS), we found a statistically significant difference for all three bolus textures ($p < 0.0001$). Analysis of variance for the post-swallowing residue in spinal onset (ALSFRS) revealed a statistically significant difference ($p < 0.0001$) for most of the comparisons between groups for all three textures. For the premature spillage in bulbar onset (b-ALSFRS), we found a statistically significant difference for all three textures ($p < 0.0001$). Analysis of variance for the post-swallowing residue in bulbar onset (b-ALSFRS) showed a statistically significant difference ($p < 0.0001$) for most of the comparisons between groups for all three textures. Kruskal–Wallis test showed a highly significant association between the classes of severity in bulbar forms and all the FEES parameters, no matter what type of bolus was administered ($p < 0.0001$), whereas a

* Corresponding author at: ENT, Audiology and Phoniatrics Unit, Department of Clinical and Experimental Medicine, University of Pisa, via Paradisa 2, I-56124 Pisa, Italy. Fax: +39 050 997501.
E-mail address: bruno.fattori@med.unipi.it (B. Fattori).

http://dx.doi.org/10.1016/j.anl.2016.07.002

significant correlation in spinal forms only for post-swallowing residue with solid ($p = 0.026$) and semisolid ($p = 0.031$) boluses.

Conclusion: There is a highly significant relationship as the following between the FEES parameters and the disease severity assessed via ALSFRS and b-ALSFRS: classes of greater severity entail a greater deterioration of FEES parameters. FEES can be considered a good indicator of the dysphagia severity and a useful test for the follow-up of dysphagia in patients with ALS, whether of spinal or bulbar onset.

1. Introduction

Amyotrophic Lateral Sclerosis (ALS), first described by Jean-Martin Charcot in 1869, is a neurodegenerative disorder characterized by upper and lower motor neuron degeneration, initiating in adulthood and leading to death within 1–5 years [1]. The phenotypic variability depends on the body region of symptoms onset (spinal onset or bulbar onset), the predominant upper or lower motor neuron involvement and the rate of disease progression. The onset of this disease is insidious, its course is inexorably progressive and prognosis is poor; the cause of death is generally respiratory insufficiency due to atrophy of the diaphragm and the intercostal muscles.

Dysphagia is one of the most frequent, invalidating and feared clinical features, possibly leading to malnutrition, *ab ingestis* pneumonia and dehydration, and it must be recognized early in clinical practice. It may be the presenting symptom in 30% of the cases and nevertheless occurs in about 80% of the cases during the course of the disease [2]. This is to be considered mainly the result of a mixed type of dysphagia because it involves both the central motor neuron (pseudo-bulbar paralysis) and the second motor neuron located in the motor nuclei of the brainstem (bulbar paralysis) [3]. Hyposthenia is therefore present both with hyperactive proprioceptive reflexes, and atrophy with weakness and twitching of the facial, tongue and pharyngeal muscles.

Dysphagia affects the first three phases of swallowing (buccal, oral and pharyngeal) [4,5] and progresses to make it impossible for the patient to feed orally. The deficit in the oral phase initially causes an increase in the amount of time required to eat a meal. There may be a deficit in the soft palate muscles, which may cause nasal reflux mainly of liquids, but this symptom is not always appreciable because of the concomitant deficit in lingual propulsion caused by paralysis of the hypoglossal nerve. Weakness of the pharyngeal and hyoid muscles can cause choking due to tracheo-bronchial aspiration.

Fiberoptic Endoscopic Evaluation of Swallowing (FEES) is currently a method of first choice in the study of swallowing disorders, for the possibility it offers particularly for directly viewing the pharyngeal phase of swallowing, allowing precise assessment of any premature spillage, post-swallowing residue in the hypo-pharyngeal region, as well as penetration and/or aspiration incidents in the lower respiratory tract. Several works in the literature compare the results from FEES investigations with those obtained with Videofluoroscopy (which is currently considered by most authors to be the gold standard for the study of swallowing), showing good specificity and sensitivity between the two tests [6–12].

Furthermore, FEES also has several advantages in the method itself, which are the following: easy to use, high tolerability, no exposure to radiation, possibility of bedside examination, repeatability and very low cost. The purpose of this study is to evaluate in a group of patients with ALS the possible relationship between the clinical severity of the disease and the main parameters of FEES, such as the following: premature spillage, post-swallowing residue (pooling amount) and tracheo-bronchial aspiration.

2. Materials and methods

We studied 202 patients with ALS (95 women and 107 men; mean age 64.68 ± 11.12 yrs SD); the onset of the disease was spinal in 136 of these and was bulbar in 66. The patients had come to our attention at the clinic for the study of dysphagia at the University Hospital of Pisa between 2012 and 2014. We classified the patients clinically according to the Amyotrophic Lateral Sclerosis Functioning Rating Scale (ALSFRS) and the b-ALSFRS subscale (bulbar scale) (Table 1) [13–15]. The ALSFRS scale is a validated questionnaire based on a score, which measures physical function while performing normal daily activities. It includes four domains (bulbar functions, gross motor tasks, fine motor tasks, and respiratory function) with scores ranging from 0 (severe disability) to 4 (no disability). Based on the overall score, the 202 patients were divided into three classes with increasing severity. According to

Table 1
ALS Functional Rating Scale, bulbar subscale (b-ALSFRS).

1. Speech	
4	Normal speech processes
3	Detectable speech disturbance
2	Intelligible with repeating
1	Speech combined with non-vocal communication
0	Loss of useful speech
2. Salivation	
4	Normal
3	Slight but definite excess of saliva in mouth; may have nighttime drooling
2	Moderately excessive saliva; may have minimal drooling
1	Marked excess of saliva with some drooling
0	Marked drooling; requires constant tissue or handkerchief
3. Swallowing	
4	Normal eating habits
3	Early eating problems – occasional choking
2	Dietary consistency changes
1	Needs supplemental tube feeding
0	NPO (exclusively parenteral or enteral feeding)

Source: Adapted from Cedarbaum et ?al. [15].

Please cite this article in press as: Fattori B, et al. Dysphagia in Amyotrophic Lateral Sclerosis: Relationships between disease progression and Fiberoptic Endoscopic Evaluation of Swallowing. Auris Nasus Larynx (2016), http://dx.doi.org/10.1016/j.anl.2016.07.002