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Birgitta Bergendal & Anita McAllister

To cite this article: Birgitta Bergendal & Anita McAllister (2017): Orofacial function and monitoring of oral care in amyotrophic lateral sclerosis, Acta Odontologica Scandinavica, DOI: [10.1080/00016357.2016.1276212](https://doi.org/10.1080/00016357.2016.1276212)

To link to this article: <http://dx.doi.org/10.1080/00016357.2016.1276212>



Published online: 12 Jan 2017.



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ORIGINAL ARTICLE

## Orofacial function and monitoring of oral care in amyotrophic lateral sclerosis

Birgitta Bergendal<sup>a,b</sup>  and Anita McAllister<sup>c,d</sup> 

<sup>a</sup>National Oral Disability Centre for Rare Disorders, The Institute for Postgraduate Dental Education, Jönköping, Sweden; <sup>b</sup>School of Health and Welfare, Jönköping University, Jönköping, Sweden; <sup>c</sup>CLINTEC, Division of Speech and Language Pathology, Karolinska Institutet, Stockholm, Sweden; <sup>d</sup>Functional Area Speech and Language Pathology, Karolinska University Hospital, Stockholm, Sweden

### ABSTRACT

**Objective:** The aim was to assess orofacial function and monitor oral care in patients with amyotrophic lateral sclerosis (ALS) to maintain oral comfort and oral health.

**Materials and methods:** A case series of 14 patients newly diagnosed with ALS accepted to participate in a quality improvement project. After initial examinations, baseline oral conditions were obtained and the patients were seen every 3 months. Nordic Orofacial Test-Screening (NOT-S) was used for evaluation of orofacial function.

**Results:** Patients were grouped according to initial symptoms in a bulbar group and a spinal group with eight and six patients, respectively. The mean age at diagnosis was 62.8 years. All were dentate with a mean of 26.7 natural teeth. Most patients had very good oral and dental conditions. As expected, orofacial functions were differently affected in the two groups; at initial NOT-S registration, the mean total score was 5.6 (range 3–8 domains) in the bulbar group and 0.7 (0–2 domains) in the spinal group. At final registration, the corresponding figures were 6.1 and 3.2. Oral and dental aids were introduced according to need.

**Conclusions:** In the bulbar group, several orofacial functions became impaired at an early stage of disease development, and at final registrations many vital orofacial functions were severely compromised. The spinal group was less severely affected orally. However, all individuals irrespective of type of initial symptoms needed assistance in performing oral hygiene measures in the latter part of the disease period. Good oral health and oral comfort could be maintained in all participants and no other dental treatment was needed.

### ARTICLE HISTORY

Received 22 June 2016  
Revised 15 November 2016  
Accepted 19 December 2016

### KEYWORDS

Amyotrophic lateral sclerosis; NOT-S; oral care; orofacial function

### Introduction

Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disease characterized by degeneration of motor neurons and progressive muscular paralysis [1]. It is a rare disorder with a prevalence of 1–9 in 100,000 with a mean age of onset of 60 years [1,2]. There are many different forms of ALS, and most cases are sporadic, while at least 10% [3] or up to 10% are familial [1]. The classic form of ALS is most common (OMIM 105400), with a male/female ratio of 1.5/1. Some patients have their first symptoms from the oral region (bulbar onset), such as difficulties to speak, chew and swallow, while others have initial problems from the limbs (spinal onset); hand, foot, arm or leg. Spinal onset is more common and seen in two of three cases [1]. Many different pathways have been described which promote disease pathology, but despite advances in identifying genes contributing to ALS pathogenesis, there is only one currently approved therapeutic drug, riluzole [4].

Common problems of speech intelligibility due to dysarthria in individuals with ALS [5] and the need for early speech therapy intervention [6,7], inefficient swallow pressure to clear the oropharynx of saliva resulting in drooling [8,9], as

well as dysphagia [9,10] have attracted much attention from researchers, whereas mostly single case reports have described more rare symptoms like masticatory muscle pain and progressive limitation of mouth opening [11] and macroglossia [12]. Most individuals with ALS acquire oral problems and difficulties to maintain good oral health, either because of impaired orofacial function or impaired function of the hands. In combination with increasing oral immobility there is an evident risk of plaque accumulation which can compromise oral health. Immobility of the tongue causes accumulation of plaque and debris that can form a thick coating on the dorsum of the tongue, which may lead to bad breath [13], and has, in frail elderly, been shown to be a risk factor for aspiration pneumonia [14]. Also, medications with dryness of the mouth as a side effect are commonly used, which increases the risk for dental caries and gingival inflammation. Compromised oral comfort and quality of life will be the result if not special attention is given to the mouth and oral functions. According to guidelines by the EFNS Task Force, management of ALS should be supportive, palliative and multidisciplinary [15,16]. Individuals who are diagnosed with ALS should ideally be treated by a multi-professional team of medical specialists, dieticians, speech- and language

pathologists (SLPs), and occupational therapists. This multi-professional approach has been shown to improve care and extend survival [17,18]. However, patients with ALS are currently monitored for changes in weight, lung capacity, speech, dysphagia, and many other functions, but it has not been customary to involve dental professionals in the team for monitoring of orofacial functions and oral health. This probably explains the very scarce amount of information available in the literature on oral health maintenance care in this vulnerable patient group. A recent case report on black hairy tongue in a patient with ALS recommended the dedicated support from a dental hygienist for the maintenance of good oral hygiene and specifically tongue hygiene [19]. However, neither a comprehensive assessment of the different aspects of orofacial dysfunction in ALS, nor recommendations on accurate domiciliary oral hygiene and proper training of personal assistants for assisted oral care have been reported earlier despite the impact of oral health on perceived quality of life [20]. The aim of our study was to monitor orofacial function and oral care in patients with ALS, in order to maintain optimal oral comfort and oral health throughout the disease period.

## Material and methods

Patients diagnosed with ALS or motor neuron disease by a neurologist were consecutively offered a referral to the National Oral Disability Centre for Rare Disorders in Jönköping, Sweden, to participate in a prospective quality improvement project on orofacial function and maintenance of oral health. Participation was voluntary and free of cost and all patients signed a consent form. Most patients were referred from the nearest hospital, but three were included from another hospital in the region. The case series of participants were divided into two groups according to their answers to a question on which were their primary symptoms leading to the diagnosis. One group described initial symptoms from the oral region or the throat, such as hoarseness, slurred speech, a numb or rigid tongue, difficulties to swallow or coughing; the bulbar group. The other group described initial symptoms from the limbs, such as numbness or weakness in thumbs, hands, feet or legs; the spinal group.

### Oral and dental examinations

At the first appointment an overall assessment was made on oral health status and oral hygiene routines by a dental team with a dentist, a dental hygienist and a dental nurse/co-ordinator. Clinical data were based on clinical examination and panoramic radiography.

### Assessment of orofacial function

Orofacial function was assessed using the Nordic Orofacial Test-Screening (NOT-S), which consists of a structured interview and a clinical examination [20]. The interview includes six domains: I Sensory function, II Breathing, III Habits, IV

Chewing and swallowing, V Drooling and VI Dryness of the mouth, and the examination includes the six domains: 1 Face at rest, 2 Nose breathing, 3 Facial expression, 4 Masticatory muscle and jaw function, 5 Oral motor function and 6 Speech. Most domains include more than one question/assessment depending on complexity, and one or more 'yes' responses for impairment in a domain equals a score of one point. The maximum score in the test is thus 12 points. The screening is easy to perform without other equipment than a picture manual. The time for a NOT-S screening varies from 5 to 13 min [21]. The examination form is available in many languages and can be downloaded at [www.mun-h-center.se](http://www.mun-h-center.se). For visualization of type of orofacial impairment and to make comparisons between groups with different diagnoses easier, the NOT-S scores have been presented as dysfunction profiles [22]. Assessment of orofacial function was made at the first dental visit after diagnosis, and the patients were thereafter offered encounters with the dental team with around 3-month intervals. When a total NOT-S score of six points or more was registered twice, indicating severely compromised orofacial function, no more registrations were made. Data from the initial and the final registrations were used for compiling of results.

### Oral and dental care

Professional tooth cleaning was performed and advice on hygiene devices during regular tooth cleaning was given at the first visit. The objective was to reach a base-line status of good oral health, and to maintain that during the disease period of progressing degenerative changes in oral and/or hand function. Additional oral aids for assisted tooth brushing, cleaning of the tongue, reduction of bad breath, and improving drinking and eating were introduced according to developing needs.

## Results

### Included patients

Fourteen patients with ALS were monitored for oral health and orofacial function. They were nine women and five men, with a mean age at the time of diagnosis of 62.8 years. The bulbar group comprised eight individuals, five women and three men, with a mean age of 64.1 years (range 54–79). In the spinal group, there were six individuals, four women and two men with a mean age of 61.0 years (range 49–73). The mean time for all patients from diagnosis to when deceased was 2 years 4 months (range 9 months to 6 years 11 months). For the bulbar group, the mean time was 1 year 10 months (range 9–32 months) and for the spinal group 3 years (range 18–83 months). Eight individuals were deceased within 2 years (9 to 24 months) after diagnosis, six had initial oral/bulbar symptoms. One male individual with limb symptoms lived with the disease for 6 years 11 months until deceased due to respiratory failure. He had no oral symptoms during the 3 years and 10 months he was monitored in the study.

**Table 1.** Dental status in 14 individuals with ALS.

Patients	Teeth in maxilla		Teeth in mandible		Total no. of teeth		Fixed dental prostheses		Pontics		Root filled teeth	
	N	Range	n	Range	n	Range	n	Range	n	Range	n	Range
Bulbar group	12.6	11–14	13.0	10–15	25.6	22–30	4.1	0–10	0.6	0–2	3.5	0–8
Spinal group	14.3	13–16	13.8	12–16	28.2	26–30	2.7	0–8	0.5	0–2	1.2	0–5
All	13.6	11–16	13.6	10–16	26.7	22–30	3.5	0–10	0.6	0–2	2.5	0–8

### Oral and dental status

All patients were dentate with bilaterally stable occlusion, and 12 had good oral health and dental status at the first examination. One patient with a severely compromised dentition had a history of dental fear and avoidance of dental treatment, and one individual was a heavy smoker and had a medium level of marginal periodontitis. The mean number of teeth in both jaws was 26.7 (range 22–30), 13.6 in each of the jaws (Table 1). Ten patients had fixed dental prostheses (FDPs) with crown therapy on 49 teeth (mean 3.5, range 0–10). Five patients had in all eight pontics (mean 0.6, range 0–2). There were 35 root filled teeth in 10 individuals (mean 2.5, range 0–8). The need for dental treatment in the group was low, only four were in need of some dental intervention. Two individuals had a need for dental fillings due to mild dental caries in 1 or 2 teeth, respectively. The patient with dental fear had four root remnants and extensive caries and was in need of dental treatment under general anaesthesia. The patient with marginal bone loss was treated with scaling and depuration, but had no need for surgery.

### Oral and dental care

No individual had a need for dental treatment after the interventions identified at the initial examination were carried out. The oral situation of good dental status and oral comfort at the start of the monitoring period could be maintained until they were deceased. Most patients used an electric tooth brush. All individuals needed help from a family member or a personal assistant to perform oral hygiene measures towards the end of their disease period.

### Tongue changes

In the later part of the disease period most patients suffered from oral inactivity, which caused retention of mucous and viscous saliva and debris, and thick coating of surface plaque and elongation of filiform papillae on the dorsal surface of the tongue. Partial muscle atrophy in a few cases caused grooves and bumps on the surface of the tongue, but fasciculations or severe macroglossia were not observed (Figure 1).

### Use of oral aids

In addition to devices for tooth cleaning, other oral aids were introduced when dental hygiene measures were carried out by others. A dental shield was recommended to stabilize the lower jaw and relieve fatigue in jaw muscles, and also to

protect the fingers of the care giver. Cheek retractors were used to facilitate tooth cleaning and get good access and a good view of the teeth. Most individuals were introduced to use a tongue scraper. To get access to the dorsum, the tip of the tongue was pulled out and firmly held with gauze while plaque and debris was removed. Oral swabs were used to moisturize the oral cavity, to remove food remnants and to add fluoride solution and/or fluid to minimize bad breath. A straw for drinking with a valve preventing reflux of water or other drinks back into the glass was much appreciated (Figure 2).

At the encounters with the dental team, soft cushions were often used to minimize pressure and help to obtain a relaxed and comfortable position in the dental chair when professional tooth cleaning was performed. Many patients were treated sitting in their wheel chairs if only examination and instruction in the use of oral aids were carried out.

### Communication aids

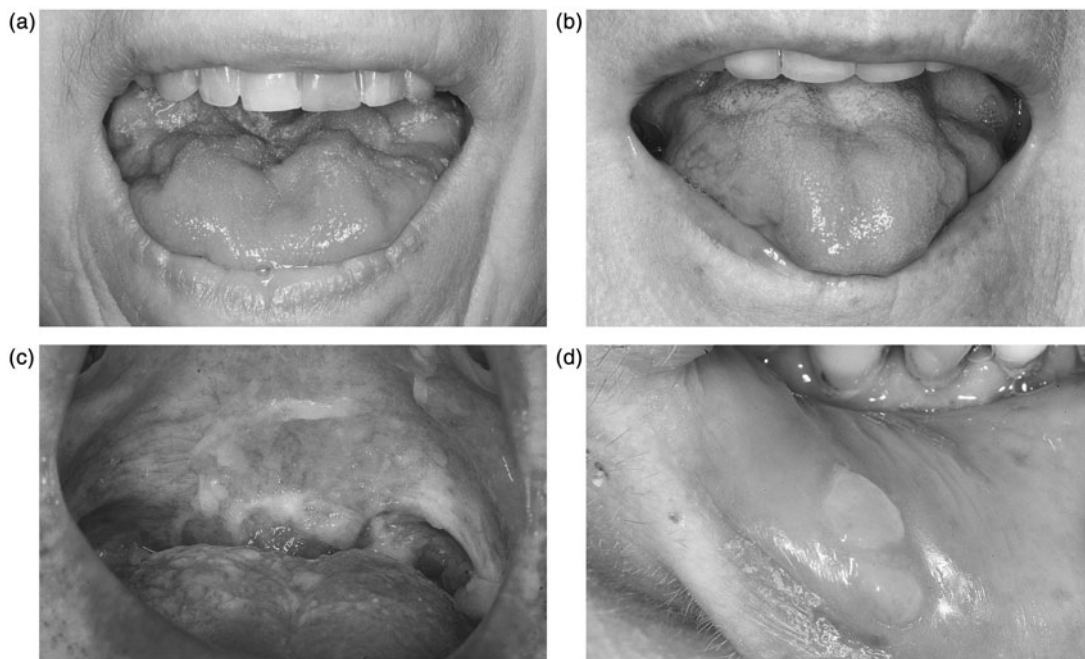
Most patients in the bulbar group had various forms of augmentative and alternative communication (AAC) prescribed by the SLP. Only one patient used high-tech, computerized aids to communicate. The use of these aids in everyday situations varied. During the visits to the dental clinic, the patients most often preferred face-to-face conversation to indicate basic wants or needs and used low tech AAC aids such as gestures, hand writing, or alphabet boards to mark agreement or disagreement when answering questions on symptoms, oral care routines and the use of oral aids. For patients in the bulbar group, this prevented communicative initiatives, especially towards the later stages of the disease.

### Orofacial function

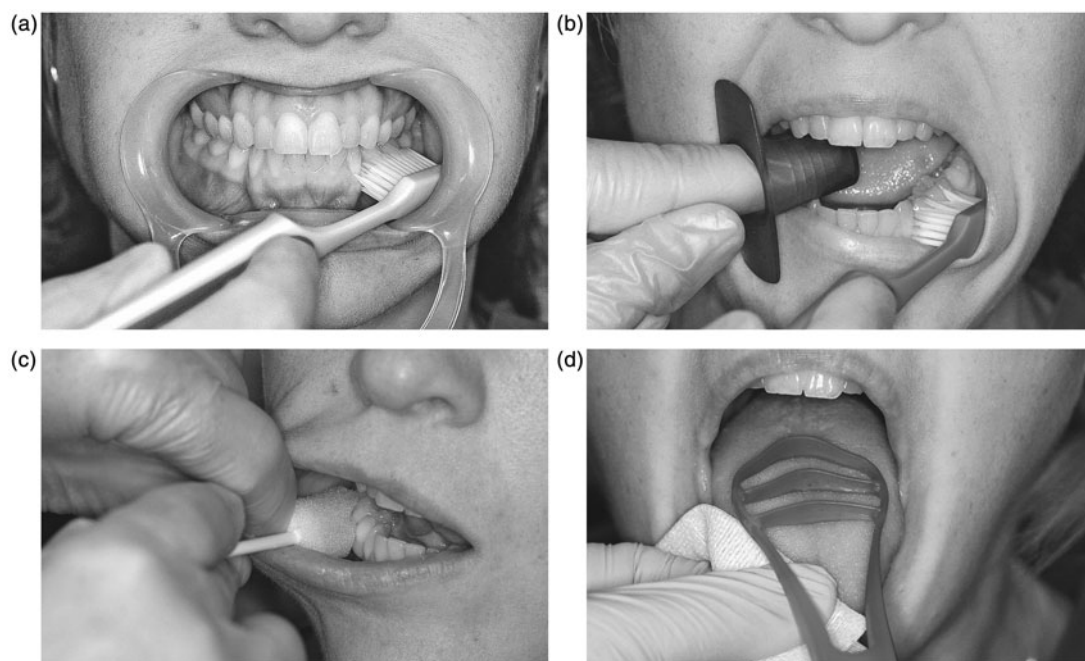
The total NOT-S scores for the two groups are compiled in Table 2. In all, there were 58 NOT-S registrations (mean 4.1, range 2–13 per individual). The total number of registrations in the bulbar group was 21 (mean 2.6, range 2–4 per individual), and in the spinal group 37 registrations (mean 6.2, range 2–13 per individual).

In the initial NOT-S registrations, the mean total score for the bulbar group was 5.6 (range 3–8) and for the spinal group 0.7 (range 0–2). In the final NOT-S registration, the mean total score for the bulbar group was 6.1 (range 3–8) and for the spinal group 3.2 (range 0–6).

The NOT-S registrations per domain are shown in Table 3. The most prevalent domains affected on the first NOT-S registrations in the bulbar group were IV Chewing and



**Figure 1.** Oral signs in four patients with ALS. (a) Tongue with typical grooves from partial muscular atrophy. (b) Tongue with surface coating and partial muscular atrophy. (c) Soft palate with debris and plaque from muscular inactivity of tongue and palate. (d) Bite wound on lower lip.



**Figure 2.** Oral aids for assisted mouth and tooth cleaning. (a) Cheek retractors to facilitate access to the mouth and teeth. (b) Dental shield for bite support and stabilization of the lower jaw, and relaxation of jaw muscles. (c) Oral swab to clean vestibulum, moisturize the oral mucosa or apply fluoride solution and fluid to cure bad breath. (d) Tongue scraper to reduce plaque and debris, tip of the tongue held out using gauze.

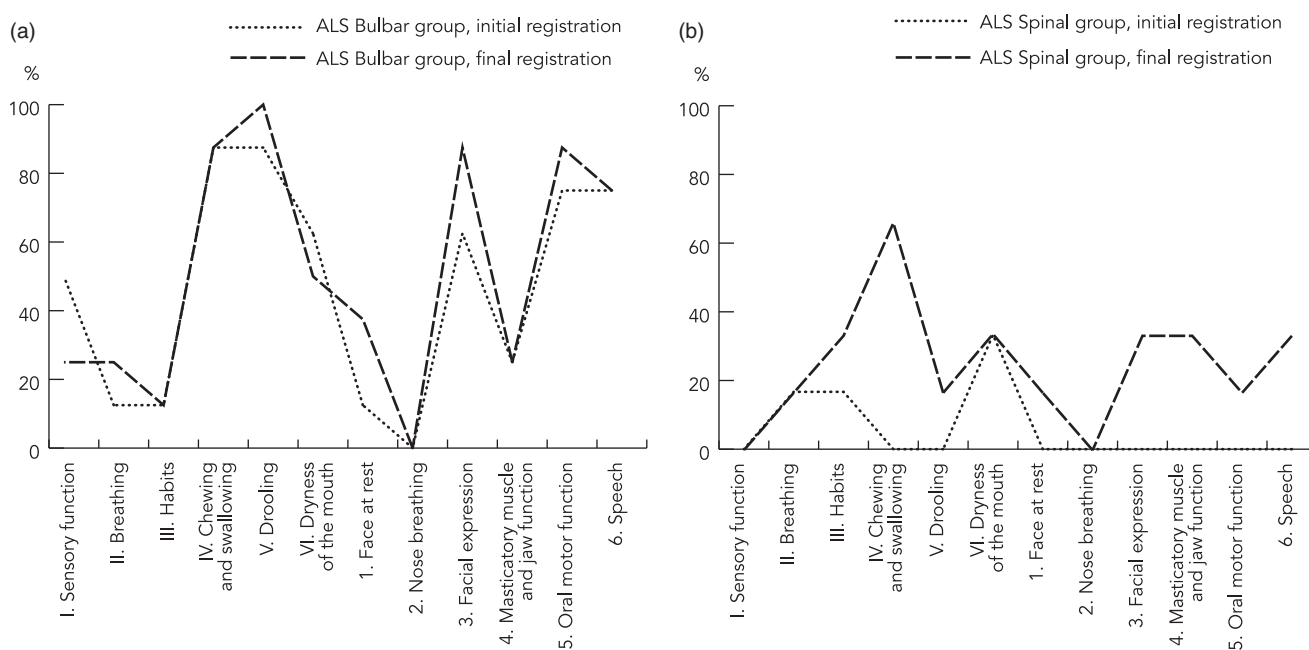
**Table 2.** Distribution of NOT-S results for orofacial function in 14 individuals, 9 women and 5 men, with ALS at initial and final registrations.

Patients	Number and gender in examined individuals			No. of NOT-S registrations		NOT-S initial total scores		NOT-S final total scores	
	Female	Male	All	Mean	Range	Mean	Range	Mean	Range
Bulbar group	5	3	8	2.6	2–4	5.6	3–8	6.1	3–8
Spinal group	4	2	6	6.2	2–13 <sup>a</sup>	0.7	0–2	3.2	0–6
All	9	5	14	4.1	2–13	3.5	0–8	4.9	0–8

<sup>a</sup>One individual in the limb group was examined 13 times and had a total score of 0 in all of them.

**Table 3.** Relative distribution of points per domain on NOT-S in individuals with ALS at initial and final registrations.

Domains of NOT-S	NOT-S initial registration				NOT-S final registration			
	Bulbar group (n = 8)		Spinal group (n = 6)		Bulbar group (n = 8)		Spinal group (n = 6)	
	n	%	n	%	n	%	n	%
<b>NOT-S interview</b>								
I Sensory function	4	50.0	0	0	2	25.0	0	0
II Breathing	1	12.5	1	16.7	2	25.0	1	16.7
III Habits	1	12.5	1	16.7	1	12.5	2	33.3
IV Chewing and swallowing	7	87.5	0	0	7	87.5	4	66.7
V Drooling	7	87.5	0	0	8	100.0	1	16.7
VI Dryness of the mouth	5	62.5	2	33.3	4	50.0	2	33.3
<b>NOT-S examination</b>								
1 Face at rest	1	12.5	0	0	3	37.5	1	16.7
2 Nose breathing	0	0	0	0	0	0	0	0
3 Facial expression	5	62.5	0	0	7	87.5	2	33.3
4 Masticatory muscle and jaw function	2	25.0	0	0	2	25.0	2	33.3
5 Oral motor function	6	75.0	0	0	7	87.5	1	16.7
6 Speech	6	75.0	0	0	6	75.0	2	33.3


**Figure 3.** Orofacial dysfunction profiles based on frequencies of NOT-S domain scores (%) in individuals with ALS using the NOT-S. (a) Dysfunction profiles at initial and final NOT-S registrations in the bulbar group (n = 8). (b) Dysfunction profiles at initial and final NOT-S registrations in the spinal group (n = 6).

swallowing (87.5%), V Drooling (87.5%) and VI Dryness of the mouth (62.5%) in the NOT-S interview, and 3 Facial expression (62.5%), 5 Oral motor function (75%) and 6 Speech (75%) in the NOT-S examination. In the spinal group two patients (33.3%) had a score in the domain VI Dryness of the mouth, besides that there were only one score each in the domains II Breathing and III Habits, respectively, in the interview. The spinal group had no scores on the NOT-S examination at the initial visit.

In the final registration, the most prevalent domains in the bulbar group in the NOT-S interview, found in almost all patients, were V Drooling (100%), IV Chewing and swallowing (87.5%) and in the NOT-S examination the most prevalent domains were 3 Facial expression (87.5%), 5 Oral motor function (87.5%) and 6 Speech (75%). In the final registration of the spinal group, domain IV Chewing and swallowing (66.7%) was the only domain where more than two patients had a

score in the NOT-S interview. In the NOT-S examination, there were still only scattered scores by one or two patients in the spinal group. Dysfunction profiles of total NOT-S scores at initial and final registrations for the two groups are shown in Figure 3.

## Discussion

ALS is a rare disorder with an incidence of around 200 individuals per year in Sweden. The number of included patients was limited, since the study was conducted at a regional hospital in a small county. However, the results are well in line with the well-known course of the disease, and unambiguously point to a need for patients with ALS to be given high priority in dental care. In order to confirm the results further studies should aim at a broader multi-centre approach.

We monitored orofacial function and oral care in 14 patients with ALS from the time they were diagnosed until they were deceased. The patients were offered to participate by the medical team at their hospital shortly after they were diagnosed, and participation was voluntary and free of cost. During the time of the study, from 2013, the Swedish Dental Insurance system allows patients with a diagnosis of ALS free dental treatment. There were more women than men in the study group, nine and five, respectively, which might be attributed to chance since the group was small and ALS is a rare disease with a low incidence rate. It could also reflect a different attitude towards dentistry, since women have been known to have more regular dental visits than men. In a US study on dental care utilization a higher percentage of women relative to men reported an annual visit, and the difference was constant from 1995 to 2008 [23]. However, in an epidemiologic study from the region, the Jönköping studies on oral health 2013 [24], all but 4–7% among 50–80 year olds reported regular dental visits and gender differences were therefore not evaluated.

The participants were grouped according to initial symptoms into a bulbar-onset group and a spinal-onset group with 8 and 6 individuals, respectively. The dental health was very good in 12 individuals, who had a history of regular dental visits and almost full dentitions. This is in accordance with the regional Swedish Jönköping epidemiologic study 2013, where a majority, 93–96% in the age cohorts 50–80 years, regularly visited a dental clinic [24]. The mean number of existing teeth was 26.7, which was slightly higher than in the regional study where the cohort of 60- and 70-year-olds had a mean of 25.0 and 22.5 existing teeth, respectively [25]. Many were treated with root fillings and prosthetic therapy (single crowns and small FDPs). In an earlier study, with material from the Jönköping studies 1993, approximately two-thirds of the 60- and 70-year olds were provided with single crowns and/or FDPs [26]. Thus, the dental status of the small group of patients with ALS reflected the dental status of the population in the region. Healthy oral conditions were maintained and no restorative needs were seen during the time of the study.

Orofacial screening with NOT-S showed more severe orofacial impairment in the bulbar group from the start of the study, which conforms to the results in a recent review that bulbar onset of ALS has consistently been associated with a poorer prognosis than spinal onset [27]. Individuals in the spinal group were followed for a mean of 8 months longer than the bulbar group, and the mean number of NOT-S registrations therefore was more than twice as many in the spinal compared to the bulbar group (mean 6.2, range 2–13 vs 2.6, range 2–4). The mean total score was 5.6 in the bulbar group and 0.7 in the spinal group at the initial examination, and 6.1 and 3.2, respectively, at the final examination. A Finnish study on diagnostics in ALS reported that people with bulbar-onset disease are more likely to experience a delayed diagnosis [7]. This might further explain the more severe orofacial symptoms at first registration and the shorter follow-up time in the bulbar group in our study. However, a recent review showed conflicting results concerning diagnostic delay [27]. Especially, patients with bulbar-onset ALS

would benefit from introduction to monitoring of oral health and speech therapy intervention early in the course of the disease.

The dysfunction profile of the bulbar group at final registration showed similarities to the profile in patients with advanced Parkinson's disease [28]. Patients with ALS are nowadays routinely monitored for weight loss, which can be caused by impaired appetite for psychological reasons as well as difficulties to chew and swallow. A close co-operation between the dental team, the SLP and a dietician is therefore important. The most common domains with scores in the bulbar group were V Drooling at both initial (87.5%) and final registrations (100%), and IV Chewing and swallowing (87.5% in both registrations). Drooling is caused by decreased tongue strength and decreased frequency of swallowing which results in accumulation of saliva [9]. In the final registration, in the spinal group, IV Chewing and Swallowing was reported by 66.7%, and only 16.7% reported V Drooling. Impaired muscle strength calls for alterations of diet to more soft food. Eating and speech problems are also affected by dryness of the mouth, which enhances motor difficulties including the ability to form a bolus and to swallow. Low muscle strength causing food retention, soft food, as well as dryness of the mouth are well known risk factors for dental diseases, but the good compliance of the patients and their care givers in combination with regular recall visits to the dental team were sufficient to prevent dental treatment needs. We used traditional methods in the maintenance care of patients with ALS and the proposed additional aids for assisted oral hygiene are commonly used in oral care in patients with special needs. The use of a tongue scraper helps reduce halitosis [13] and could in combination with good dental hygiene reduce oral plaque and help reduce the risk for aspiration pneumonia [14].

Individuals with ALS are an extremely vulnerable group where a majority will develop a situation where communication and oral functions are severely compromised, and become dependent on other persons to perform oral hygiene measures and assist in communication. They therefore are in need of special attention by a dental team both in the early stage after diagnosis to establish healthy oral conditions, and throughout the disease period to maintain oral health and prevent oral treatment interventions as well as a SLP to provide adequate AAC.

The prerequisites for our study were that it was performed in a context of dental care free of charge, and in a population where oral health in comparable age cohorts is generally very good. Based on this experience our recommendations for dental care in patients with ALS are as follows:

- Dental professionals should be represented in multidisciplinary teams for ALS.
- All individuals diagnosed with ALS should be referred to a specialized dental team.
- Oral conditions should be optimized to ensure good oral comfort and oral health as early as possible in the course of the disease.
- Regular oral maintenance care and support should be performed to maintain oral health and avoid dental disease.

- Suitable oral aids should be introduced according to need.

In conclusion, in a small group of patients with bulbar and spinal-onset ALS, we showed that orofacial functions became compromised in both groups, and that oral comfort and oral health could be maintained during the disease period with minimal clinical intervention. Practical advice and support on the use of aids for oral hygiene and comfort were the most important part of the encounters with the dental team.

## Acknowledgements

We sincerely thank the patients who participated in the study and their families. The project was initiated in memory of Ulla Carin Lindquist, founder of the Ulla Carin Lindqvist Foundation for ALS research. The valuable contribution of the team members dental hygienist Ann-Cathrin Widal, coordinator Annica Krogell and dietician Elisabet Allheim is gratefully acknowledged.

## Disclosure statement


The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.


## Notes on contributors

**Birgitta Bergendal**, is a specialist in prosthetic dentistry and former head of the National Oral Disability Centre for Rare Disorders at the Institute for Postgraduate Dental Education in Jönköping, Sweden. She initiated the study, performed the clinical registrations, and wrote the original draft of the manuscript. She worked on analysis of data and the final version of the manuscript.

**Anita McAllister**, is a speech and language pathologist employed as associate professor at CLINTEC, Karolinska Institutet and Karolinska University Hospital, Stockholm, Sweden. She worked on analysis of data, contributed to the draft, and the final version of the article.

## ORCID

Birgitta Bergendal  <http://orcid.org/0000-0002-8009-8268>

Anita McAllister  <http://orcid.org/0000-0003-2208-0630>

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