ACTA ODONTOLOGICA SCANDINAVICA
2018, VOL. 76, NO. 4, 294–298
https://doi.org/10.1080/00016357.2017.1420228

Taylor & Francis
Taylor & Francis Group

ORIGINAL ARTICLE

Oral health status of hospitalized amyotrophic lateral sclerosis patients: a single-centre observational study

Rena Nakayama[a], Akira Nishiyama[a], Chiharu Matsuda[b], Yuki Nakayama[b], Chiyoko Hakuta[c] and Masahiko Shimada[a]

[a]Orofacial Pain Management, Oral Health Sciences, Graduate School of Medical and Dental Sciences, Tokyo Medical and Dental University, Tokyo, Japan; [b]ALS Nursing Care Project, Tokyo Metropolitan Institute of Medical Science, Tokyo, Japan; [c]Oral Health Sciences for Community Welfare, Oral Health Sciences, Graduate School of Medical and Dental Sciences, Tokyo Medical and Dental University, Tokyo, Japan

ABSTRACT

Objective: To assess the intraoral conditions and oral function of patients with amyotrophic lateral sclerosis (ALS).

Material and methods: This single-centre, cross-sectional observational study included 50 ALS patients, who were treated with tracheostomy positive-pressure ventilation (TPPV) while hospitalized. The disease duration, TPPV duration, current number of teeth, number of occlusal units, number of decayed/missing/filled teeth, community periodontal index, bleeding on probing, dental calculus, maximum mouth opening, salivation rate, tongue anomalies (atrophy or hypertrophy) and tongue coating were determined for each patient. Differences in intraoral conditions according to disease duration or TPPV duration were statistically analysed.

Results: The maximum mouth opening was low in the included patients, with a mean distance of 13.7 ± 7.4 mm. Furthermore, the maximum mouth opening showed a significant negative correlation with both disease duration and TPPV duration. No statistically significant differences were found between any other intraoral parameters and disease duration or TPPV duration.

Conclusions: Severe dental disease is uncommon among hospitalized ALS patients who receive oral care by nurses; however, mouth opening is very restricted in these patients. Early intervention for restricted mouth opening, directed by a dentist or dental hygienist, is essential in this population.

ARTICLE HISTORY
Received 17 October 2017
Accepted 18 December 2017

KEYWORDS
Community periodontal index; maximum mouth opening; oral health; salivation rate; tracheostomy positive-pressure ventilation

Introduction

Amyotrophic lateral sclerosis (ALS) is a disease of unknown aetiology that involves sporadic and progressive degeneration of upper and lower motor neurons. The onset of ALS symptoms usually occurs in late middle age, and the incidence rate in Japan is 1–2.5 per 100,000 people per year. ALS is more common in men, with the incidence rate in Japanese men being 1.2–1.3 times higher than in Japanese women [1]. The mean survival time of patients with ALS is reportedly 40.6 ± 33.1 months without invasive ventilation and 49.1 months if patients are treated with tracheostomy positive-pressure ventilation (TPPV) [1,2]. The prevalence of ALS in Europe has been reported to be 2.16 per 10,000 people [3], and according to the same report, 50% of ALS patients die within 30 months after the onset of symptoms; 20% of patients who survive longer than 30 months die within 5–10 years.

The principal symptoms of ALS are limb disorders, dysarthria, dysphagia and respiratory muscle paralysis; cognitive impairment also occurs in some cases. ALS patients present with a myriad of oral and dental problems, including dysarthria, dysphagia and sialorrhea, all of which can negatively affect a patient's quality of life. Bergendal et al. classified ALS patients into bulbar and spinal groups based on the initial symptoms [4]. The initial symptoms of patients in the bulbar group affect the mouth or pharyngeal region, including voice hoarseness, unclear and/or inarticulate speech, tongue stiffness and difficulty with swallowing and coughing; the initial symptoms of patients in the spinal group affect the limbs, such as numbness and weakness. It has been reported that the spinal group also shows orofacial dysfunction from the initial stage of the disease; thus, irrespective of the ALS disease mode, there is a need to evaluate and support oral hygiene until the final stage of the disease. Although there have been reports investigating salivation [5], numbers of teeth and dental restorations in ALS patients [4], there have been no reports about caries or periodontal disease in this population. Therefore, further studies are needed in order to facilitate appropriate oral hygiene management for ALS patients, including patient and caregiver education. For these reasons, this study investigated the intraoral conditions and oral function of hospitalized ALS patients, specifically investigating which intraoral factors are associated with increased disease duration and TPPV duration.

CONTACT Akira Nishiyama anishi.tmj@tmd.ac.jp Orofacial Pain Management, Oral Health Sciences, Graduate School of Medical and Dental Sciences, Tokyo Medical and Dental University, Bunkyo-ku, Tokyo, Japan

Materials and methods

Study participants

Among ALS patients hospitalized at Sayama Neurology Hospital (Saitama Prefecture, Japan) from May to November 2014, fifty patients who were treated with tube feeding and TPPV were included in this study (31 men and 19 women). None of the included patients had a history of severe heart failure, pneumonia, liver disease or kidney disease, and their symptoms had been stabilized. The oral cavity of each patient was cleaned at least twice per day by nurses, including the removal of accumulated liquid from the oral cavity by aspiration, intratracheal aspiration, care of the tongue and mucosa, and teeth cleaning with a toothbrush. None of the included patients were undergoing treatment to reduce salivation, such as radiotherapy or botulinum toxin injection. Patients provided both verbal and written consent to participate in the study. The consent of family members was obtained for patients with limited ability to communicate.

This study was approved by the Ethics Committees of Sayama Neurology Hospital and the Tokyo Metropolitan Institute of Medical Science (approval no. 13-06).

Study details

A total of 13 parameters were evaluated, including age, sex, disease duration, TPPV duration, current number of teeth, number of occlusal units (OU), number of decayed/missing/filled teeth (DMFT), periodontal disease parameters [community periodontal index (CPI), including the presence or absence of pocket haemorrhage and the presence or absence of dental calculus], maximum mouth opening (MMO), salivation rate, the presence of tongue anomalies (atrophy or hypertrophy and the presence or absence of tongue coating.

The OU is an index to evaluate occlusal support in the molar region. In this study, the method developed by Baba et al. was used to determine the OU [6], such that a pair of occluded molars was counted as 2 OU and a pair of occluded premolars was counted as 1 OU, with a maximum total of 12 OU.

The CPI is an index that was defined by the World Health Organization (WHO) in 1997, which was a modification of the CPI of treatment needs that was developed collaboratively by the WHO and the Federation Dentaire Internationale in 1982. The CPI assessment includes the following six teeth or pairs of teeth (Federation Dentaire Internationale two-digit notation): (i) 16 and 17; (ii) 11; (iii) 26 and 27; (iv) 36 and 37; (v) 31 and (vi) 46 and 47. For each tooth, the depth of the gingival pocket was measured using a WHO-type probe, and the evaluation was coded as follows: 0: no signs; 1: haemorrhage present; 2: dental calculus present; 3: gingival pocket 4–5 mm deep and 4: gingival pocket at least 6 mm deep. For each of the pairs of molars, the higher code was taken to be representative, and the highest code among the six teeth or pairs of teeth was taken to be the individual patient's highest code.

The MMO is the distance between the incisal margins of the mandibular and maxillary incisors when the patient's mouth is forced open. In this study, the researcher used his or her fingers to open the patient's mouth as far as possible to measure the MMO.

Salivation rate is defined as the rate of saliva secretion when a patient is at rest and was measured by the cotton-roll method in this study. Before the measurement, saliva that had accumulated in the patient's oral cavity was removed by noninvasive aspiration at an aspiration pressure no higher than 20 kPa; intraoral and intratracheal aspirations were carried out in accordance with tracheal aspiration guidelines. Once the patient's respiratory state (rhythm, ventilatory volume and cough cessation) had normalized, medical cotton rolls were placed under the tongue and in the salivary gland aperture near the second maxillary molar. After 1 min, the cotton rolls were removed and the volume of saliva was measured. Intraoral outcomes were measured by two calibrated dental hygienists. The oral evaluation was carried out between 1:00 pm and 4:00 pm and at least 1 h after completion of oral cleaning and tube feeding, in order to minimize the effects of stimulating the oral cavity by these procedures and the subsequent fluid replacement period. At the time of measurement, the patients were supine without their heads being raised and with the pillow removed. The patients' sex, age, disease duration and TPPV duration were recorded from the medical records.

Statistical analysis

Descriptive statistical analysis was carried out with each of the above parameters. The correlation coefficients were determined between each of the continuous variables and disease duration and TPPV duration. In addition, for each of the binary variables, significant differences with respect to disease duration and TPPV duration were determined using the t-test. Statistical analyses were performed with SPSS software version 21.0 (IBM Japan, Tokyo, Japan), and $p < .05$ was considered statistically significant.

Results

The descriptive statistics are shown in Table 1. Results for patients of approximately the same ages taken from the Survey of Dental Diseases conducted by the Japanese Ministry of Health, Labour, and Welfare in 2016 were used as reference values. The mean disease duration and TPPV duration for ALS patients were 8.8 ± 5.0 and 6.3 ± 4.0 years, respectively. The mean MMO was 13.7 mm. The incidence of tongue anomalies (atrophy or hypertrophy) and tongue coatings were 60.0 and 52.0%, respectively.

There was a significant correlation between MMO and disease duration and between MMO and TPPV duration (Table 2). No statistically significant differences were found in the incidence of bleeding, dental calculus, tongue anomaly or tongue coating with respect to disease duration and TPPV duration (Table 3).

For CPI, code 2 (dental calculus present) was the most frequent at 48.0% followed by code 0 (no signs) at 30.0% (Table 4).

Table 1. Patient characteristics and results of each outcome.

	Number of patients (SD)	Reference value[a] (70–74 y)
Mean age, years (SD)	70.7 (7.0)	–
Gender (male/female), *N* (%)	31 (62.0)/19 (38.0)	–
Mean disease duration, years (SD)	8.8 (5.0)	–
Mean TPPV duration, years (SD)	6.3 (4.0)	–
Mean tooth number, *N* (SD)	23.6 (5.6)	17.3 (9.8)
Mean OU: *N* (SD)	7.5 (4.4)	–
DMFT, *N* (SD)	13.0 (6.4)	21.1 (6.4)
Bleeding, *N* (%)	21 (42.0)	38.8
Dental calculus, *N* (%)	26 (52.0)	–
MMO, mm (SD)	13.7 (7.4)	45–55
Salivation rate, mL/min (SD)	0.72 (0.6)	0.3–0.4
Tongue atrophy or hypertrophy, *N* (%)	30 (60.0)	–
Tongue coating, *N* (%)	26 (52.0)	–

[a]Results of survey of dental diseases in 2016 by the Japanese ministry of health, labour and welfare.
SD: standard deviation; TPPV: tracheostomy positive-pressure ventilation; OU: occlusal units; DMFT: decayed/missing/filled teeth; MMO: maximum mouth opening.

Table 2. Correlation coefficient to disease and TPPV duration.

	Disease duration, correlation coefficient (*p*)	TPPV duration, correlation coefficient (*p*)
Tooth number	−.024 (.866)	−.037 (.801)
OU	−.088 (.541)	−.152 (.291)
DMFT	−.024 (.867)	.012 (.933)
CPI	.063 (.622)	.031 (.833)
MMO	−.356* (.011)	−.285* (.045)
Salivation rate	.146 (.316)	.078 (.594)

TPPV: tracheostomy positive pressure ventilation; OU: occlusal units; DMFT: decayed/missing/filled teeth; CPI: community periodontal index; MMO: maximum mouth opening; $^{*}p < .005$.

Discussion

Symptoms progress rapidly after onset of ALS, and death occurs 2–5 years after onset unless an artificial respirator is fitted [3,7]. In a survey of 27 ALS patients by Hirano et al., the mean disease duration since ALS onset was 8.4 ± 5.4 years (4.4 ± 3.7 years after using invasive mechanical ventilation) [8]. In a study by Nakane et al., the mean disease duration was 5.6 ± 3.1 years [9]. These results demonstrate that some Japanese ALS patients have considerably longer survival times than the global average. Moreover, the mean disease duration was 8.8 years in this study, which is relatively long.

In a retrospective study of ALS patients by Burkhardt et al., 72 of 74 patients died of respiratory failure from one of the following causes: 15 patients had aspiration pneumonia, 23 had bronchial pneumonia, eight had concomitant aspiration and bronchial pneumonia, 20 had hypoxia and six had pulmonary embolism. In addition, it has been shown that management by noninvasive ventilation increases the risk of death due to bronchial pneumonia [10]. The proportion of ALS patients treated with TPPV reportedly differs between countries, with 24.5% in Japan [11], 21% in Korea, 2% in the northern USA and 1.5% in Canada [12]. The high rate of TPPV use in Japan is likely due to the coverage of this treatment by the national health insurance system. All patients in this study were treated with TPPV, which may explain the relatively long disease duration, the stabilization of symptoms and the relatively low incidence of bronchial pneumonia.

A comparison of the results of this study with the survey of dental diseases (2016) shows that both DMFT and CPI scores were more favourable in this study. In particular, a large proportion of patients in this study showed no signs of gingival disease. Furthermore, the salivation rate at rest was above average in this study. In general, the salivation rate at rest is considered to be 0.3–0.4 mL/min [13–15]. In a study by Suzuki et al. that examined 119 patients with halitosis, the mean salivation rate at rest was 0.13 mL/min [16]. In a survey by Muddugangadhar et al., the salivation rates were found to be 0.34 ± 0.01 mL/min in people aged 45 years and younger and 0.33 ± 0.01 mL/min in people aged 46–60 years [17]. In this study, the salivation rate at rest was higher than in healthy people or patients with diseases other than ALS. This difference may be due to abnormalities of autonomous nervous activity and other central regulatory functions [18,19]. High salivation rates have the potential to maintain intraoral buffering, and the bactericidal activity of saliva may suppress the progression of caries and gingival disease. In addition, favourable intraoral conditions may reduce the risk of aspiration pneumonia and other infectious diseases.

Although ALS patients may benefit from increased salivation, mouth opening is significantly restricted in this population. In healthy people, the MMO is typically 45–55 mm [20]. The MMO of ALS patients has previously been reported to be 20 mm [21], however, the mean MMO of patients in this study was only 13.7 mm. Restricted mouth opening places a significant burden on caregivers with respect to intraoral aspiration and oral care. One condition that gives rise to problems with mouth-opening is temporomandibular disorder (TMD) [22,23], and one proposed treatment for this condition is an exercise that forces the mouth open [24]. Insufficient data are available about whole-body management of locomotory organ dysfunction and the prevention of symptom progression in ALS patients [25,26]; however, Lui et al. have reported the efficacy of exercise therapy [27]. It is, therefore, probable that vigorous treatment of mouth opening disorders from the initial stages of the disease would effectively maintain mouth opening or at least minimize the reduction in MMO. Mouth-opening exercises are effective for increasing the range of motion of the temporomandibular joint and for stretching the masticatory muscles, especially the jaw-closing muscles. However, depending upon the statuses of residual bacteria and gingival tissues in individual patients, the mouth-opening exercise regimen must be customized in terms of the frequency of the exercise and the load duration. Thus, proactive intervention by dentists and dental hygienists will be essential from the initial stage of ALS to minimize reductions in MMO and to support oral management.

The initial symptoms can manifest differently among patients with ALS [2]. However, irrespective of the initial symptoms, it is clear that MMO decreases with increasing disease duration; therefore, it is important for dentists and dental hygienists to be proactive in educating patients and caregivers as early as possible about mouth-opening exercises and oral care.

Table 3. Relationships between binary variables and disease duration and TPPV duration.

	Disease duration, mean years (SD)			TPPV duration, mean years (SD)		
	Negative	Positive	*p*	Negative	Positive	*p*
Bleeding	8.34 (4.66)	9.42 (5.51)	.458	5.75 (3.30)	6.94 (4.82)	.333
Dental calculus	8.56 (5.19)	9.02 (4.92)	.750	6.20 (4.46)	6.30 (3.64)	.934
Tongue atrophy or hypertrophy	7.93 (5.11)	9.37 (4.94)	.326	5.52 (4.07)	6.75 (3.97)	.296
Tongue coating	8.12 (4.53)	9.42 (5.43)	.363	5.53 (3.60)	6.92 (4.32)	.224

TPPV: tracheostomy positive-pressure ventilation; SD: standard deviation.

Table 4. Numbers and proportions of patients with each CPI code for each tooth or pair of teeth.

	Block (FDI): *N* (%)							
Code	16 or 17	11	26 or 27	36 or 37	31	46 or 47	Maximum code	Reference value[a] (70–74 y)
0	12 (24.0)	16 (32.0)	14 (28.0)	13 (26.0)	12 (24.0)	12 (24.0)	15 (30.0)	9.7
1	15 (30.0)	16 (32.0)	14 (28.0)	10 (20.0)	15 (30.0)	11 (22.0)	9 (18.0)	10.6
2	13 (26.0)	12 (24.0)	15 (30.0)	16 (32.0)	19 (38.0)	17 (34.0)	24 (48.0)	20.6
3	0	0	1 (2.0)	2 (4.0)	1 (2.0)	2 (4.0)	2 (4.0)	29.0
4	0	0	0	0	0	0	0	13.8
All target teeth absent	10 (20.0)	6 (12.0)	6 (12.0)	9 (18.0)	3 (6.0)	8 (16.0)	0	16.3

CPI: community periodontal index; FDI: Federation Dentaire Internationale.
[a]Results of survey of dental diseases in 2016 by the Japanese ministry of health, labour and welfare.

It will also be beneficial to continue collecting data from the patients in this study and to evaluate outcome differences between medical institutions where different instructions about oral hygiene are given. Further studies are also needed to investigate the effects of intervention by dental experts in patients with ALS. In addition, better approaches to instructing caregivers about effective mouth-opening exercises need to be developed.

In conclusion, this study has shown that, although severe dental disease is uncommon in hospitalized ALS patients receiving oral care by nurses, such patients tend to show marked restriction of mouth opening. Therefore, dentists and dental hygienists should begin instructing ALS patients and caregivers about mouth-opening exercises from an early stage.

Acknowledgements

The authors would like to thank Dr. Takaya Numayama (director, Sayama Neurology Hospital), Dr. Masato Taira (Tokyo Medical and Dental University) and the research staff for their invaluable contributions to this work. In addition, we thank the patients who participated in this study. We would like to thank Editage (www.editage.jp) for English language editing.

Disclosure statement

The authors report no conflicts of interest.

Funding

This work was supported by Grants-in-Aid for Scientific Research by the Japan Society for the Promotion of Science (JSPS KAKENHI grant number JP.25463459).

References

[1] Amyotrophic lateral sclerosis (ALS) [Internet]. Japan intractable diseases information center. [cited 2017 Sep 3]. Available from: http://www.nanbyou.or.jp/entry/52

[2] Doi Y, Atsuta N, Sobue G, et al. Prevalence and incidence of amyotrophic lateral sclerosis in Japan. J Epidemiol. 2014;24:494–499.

[3] Kiernan MC, Vucic S, Cheah BC, et al. Amyotrophic lateral sclerosis. Lancet. 2011;377:942–955.

[4] Bergendal B, McAllister A. Orofacial function and monitoring of oral care in amyotrophic lateral sclerosis. Acta Odontol Scand. 2017;75:179–185.

[5] Mcgeachan AJ, Hobson EV, Al-Chalabi A, et al. A multicentre evaluation of oropharyngeal secretion management practices in amyotrophic lateral sclerosis. Amyotroph Lateral Scler Frontotemporal Degener. 2017;18:1–9.

[6] Baba K, Igarashi Y, Nishiyama A, et al. Patterns of missing occlusal units and oral health-related quality of life in SDA patients. J Oral Rehabil. 2008;35:621–628.

[7] Watanabe H, Atsuta N, Nakamura R, et al. Factors affecting longitudinal functional decline and survival in amyotrophic lateral sclerosis patients. Amyotroph Lateral Scler Frontotemporal Degener. 2015;16:230–236.

[8] Hirano YM, Yamazaki Y, Shimizu J, et al. Ventilator dependence and expressions of need: a study of patients with amyotrophic lateral sclerosis in Japan. Soc Sci Med. 2006;62:1403–1413.

[9] Nakane S, Izumi Y, Oda M, et al. A potential link between amyotrophic lateral sclerosis and bullous pemphigoid: six new cases and a systematic review of the literature. Intern Med. 2016;55:1985–1990.

[10] Burkhardt C, Neuwirth C, Sommacal A, et al. Is survival improved by the use of NIV and PEG in amyotrophic lateral sclerosis (ALS)? A post-mortem study of 80 ALS patients. PLoS One. 2017;12:e0177555.

[11] Borasio GD, Gelinas DF, Yanagisawa N. Mechanical ventilation in amyotrophic lateral sclerosis: a cross-cultural perspective. J Neurol. 1998;245:7–12.

[12] Kimura F. Tracheostomy and invasive mechanical ventilation in amyotrophic lateral sclerosis: decision-making factors and survival analysis. Clin Neurol. 2016;56:241–247.

[13] Dawes C. Physiological factors affecting salivary flow rate, oral sugar clearance, and the sensation of dry mouth in man. J Dent Res. 1987;66:648–653.

[14] Funayama S, Ito K, Nohno K, et al. Comparative study of salivary secretion in cotton roll method and spitting method. Niigata Dent J. 2008;38:37–43.

[15] Bourdiol P, Mioche L, Monier S. Effect of age on salivary flow obtained under feeding and non-feeding conditions. J Oral Rehabil. 2004;31:445–452.

[16] Suzuki N, Fujimoto A, Yoneda M, et al. Resting salivary flow independently associated with oral malodor. BMC Oral Health. 2016;17:23.

[17] Muddugangadhar BC, Sangur R, Rudraprasad IV, et al. A clinical study to compare between resting and stimulated whole salivary flow rate and pH before and after complete denture placement in different age groups. J Indian Prosthodont Soc. 2015;15: 356–366.

[18] Shimizu T, Hayashi H, Kato S, et al. Circulatory collapse and sudden death in respirator-dependent amyotrophic lateral sclerosis. J Neurol Sci. 1994;124:45–55.

[19] Shimizu T. Sympathetic hyperactivity and sympathovagal imbalance in amyotrophic lateral sclerosis. Eur Neurol Rev. 2013;8:46–50.

[20] Tsukahara H, Yoda T, Sakamoto I, et al. A statistical evaluation of normal maximal mouth opening of Japanese adults. Jpn J Oral Maxillofac Surg. 1998;44:159–167.

[21] Pang KM, Park JW. Masticatory muscle pain and progressive mouth opening limitation caused by amyotrophic lateral sclerosis: a case report. J Oral Facial Pain Headache. 2015;29:91–96.

[22] Okeson JP. Orofacial pain: guidelines for assessment, diagnosis and management. Chicago (IL): Quintessence; 1996.

[23] Dworkin SF, Huggins KH, LeResche L, et al. Epidemiology of signs and symptoms in temporomandibular disorders: clinical sign in cases and controls. J Am Dent Assoc. 1990;120:273–281.

[24] Yuasa H, Kino K, Kubota E, et al. The evidence-based clinical practice guidelines task force. primary treatment of temporomandibular disorders: the Japanese society for the temporomandibular joint evidence-based clinical practice guidelines, 2nd edition. Jpn Dent Sci Rev. 2013;43:89–98.

[25] Plowman EK. Is there a role for exercise in the management of bulbar dysfunction in amyotrophic lateral sclerosis? J Speech Lang Hear Res. 2015;58:1151–1166.

[26] Dal Bello-Haas V, Florence JM. Therapeutic exercise for people with amyotrophic lateral sclerosis or motor neuron disease. Cochrane Database Syst Rev. 2013;16:CD005229.

[27] Lui AJ, Byl NN. A systematic review of the effect of moderate intensity exercise on function and disease progression in amyotrophic lateral sclerosis. J Neurol Phys Ther. 2009;33: 68–87.