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Correlation between Oro-buccal Symptoms and Respiratory Function in Patients with ALS

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Introduction
Amyotrophic lateral sclerosis (ALS) is progressive neurodegenerative disorder. Subtypes of ALS with initial exclusive bulbar or spinal involvement are associated with prognosis. Predicting when respiratory failure will occur in the patient with ALS is important to plan appropriate clinical interventions. In patients with predominant bulbar weakness, it is limited to measure pulmonary function accurately. The aim of this study is to investigate correlation between oro-buccal symptoms (dysphagia, dysarthria, and sialorrhea) and respiratory function in patients with ALS according to bulbar weakness.

Methods
Medical records of 91 patients with ALS were reviewed and this is a cross-sectional study. Of these, 37 had bulbar-onset ALS (bALS) and 54 spinal-onset ALS (sALS). Functional status of subjects was scored using Korean version of Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised (K-ALSFRS-R) and especially oro-buccal symptoms were assessed using bulbar domain of K-ALSFRS-R (b-K-ALSFRS-R), which consists of speech, salivation and swallowing. Respiratory dysfunction was assessed by pulmonary function test (PFT) and diaphragm fluoroscopy. Parameters were measured as forced vital capacity (FVC), peak cough flow (PCF), maximal inspiratory pressure (MIP) and maximal expiratory pressure (MEP) in supine and sitting position. To assess pulmonary function in a non-oral way, movements of diaphragm while forced respiration were measured in centimeters by fluoroscopy. Pearson correlation analysis was used.

Results
The mean age of total 91 ALS subjects was 59 ± 10.51, 37 bALS subjects was 63 ± 8.1 years and 54 sALS patients was 57 ± 11.2 years. The mean K-ALSFRS-R and b-K-ALSFRS-R of total ALS patients was 17 ± 11.4 and 4 ± 2.47, those of bALS patients was 16 ± 11.9 and 3 ± 2.2, and those of sALS patients' was 19 ± 11.3 and 5 ± 2.3 in order. In 91 ALS patients, b-K-ALSFRS-R is statistically associated with all parameters of PFT ($p<0.05$). There was the positive linear correlation between b-K-ALSFRS-R and the left forced diaphragm movement ($p<0.05$). In sALS group, the b-ALS-FRS-R was statistically associated with supine FVC, supine MEP, sitting PCF, and sitting MEP ($p<0.05$). Among subjects with bALS, there was no statistically correlation between the b-ALS-FRS-R and any parameter of PFT and diaphragm movements.

Conclusion
This study suggest that oro-buccal symptoms could be predictors for respiratory insufficiency in ALS. In sALS patients revealed correlation with maximal expiratory

pressure, trunk muscle strength, in supine and sitting position in sALS patients. In generally sALS patients undergo oro-buccal and respiratory dysfunction in more progressed state than bALS patients. Prediction and correlation between oro-buccal symptoms and respiratory insufficiency is limited in bALS patients, therefore assessing symptoms of respiratory insufficiency such as dyspnea and orthopnea are important.

Table 1. Demographic and clinical characteristics of subjects

Variables	Total (n=91)	Bulbar-onset (n=37)	Spinal-onset (n=54)	*p*-value
Age (years)	59 ± 10.51	63 ± 8.1	57 ± 11.2	<0.01
Gender				
Male	60 (65.9%)	23 (62.2%)	37 (68.5%)	2.08
Female	31 (34.1%)	14 (37.8%)	17 (31.5%)	
Duration of disease (years)	2 ± 1.78	3 ± 1.8	1.5 ± 1.2	0.06
K-ALSFRS-R	17 ± 11.4	16 ± 11.9	19 ± 11.3	0.23
b-K-ALSFRS-R	4 ± 2.47	3 ± 2.2	5 ± 2.3	1.18

K-ALSFRS-R; Korean version amyotrophic lateral sclerosis functional rating scale revised, b-K-ALSFRS-R; bulbar domain of K-ALSFRS-R

Table 2. Pearson correlation coefficients (r) between bulbar domain of K-ALSFRS-R with each variable

Variables	Total (n=91)		Bulbar onset (n=37)		Limb onset (n=54)	
	r	*p*-value	*r*	*p*-value	*r*	*p*-value
Supine						
FVC	.290*	.005	.169	.319	.270*	.048
PCF	.265*	.011	.012	.945	.251	.067
MIP	.245*	.019	.176	.296	.190	.168
MEP	.320*	.002	.187	.269	.330*	.015
FMRD	.114	.280	-.226	.179	.113	.414
FMLD	.176	.096	-.197	.250	.139	.316
FMBD	.160	.131	-.223	.184	.141	.309
Sitting						
FVC	.247*	.018	.166	.327	.203	.141
PCF	.295*	.005	.073	.667	.291*	.032
MIP	.253*	.015	.179	.290	.207	.133
MEP	.310*	.003	.217	.197	.374*	.005
FMRD	.129	.222	-.093	.184	.162	.240
FMLD	.255*	.015	.030	.859	.244	.075
FMBD	.217*	.039	-.034	.841	.230	.095

*$p<0.05$, K-ALSFRS-R; Korean version amyotrophic lateral sclerosis functional rating scale revised, FVC; forced vital capacity, PCF; peak cough flow, MIP; maximal inspiratory pressure, MEP; maximal expiratory pressure, FMRD; the forced movement of the right diaphragm, FMLD; the forced movement of the left diaphragm, FMBD; mean value between the forced movement of both diaphragms