

신경근육재활 및 전기진단

게시일시 및 장소 : 10 월 18 일(금) 08:30-12:20 Room G(3F)

질의응답 일시 및 장소 : 10 월 18 일(금) 10:28-10:32 Room G(3F)

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Relationship between tongue pressure and functional measures in spinal and bulbar muscular atrophy

Min-Gu Kang^{1*}, Yu-Sun Min^{1†}, Dae-won Gwak¹, Jin-Sung Park², Hee-Jin Cho², Jae-Eun Lee¹, Jae-Gyeong Jeong¹

Kyungpook National University Hospital, Department of Rehabilitation Medicine¹,
Kyungpook National University Hospital, Department of Neurology²

Background/Objective

Spinal and bulbar muscular atrophy (Kennedy disease, KD) is an X-linked neurodegenerative disease which is caused by an expansion of CAG repeat sequence in the first exon of the androgen receptor gene. The disease is characterized by slowly progressive proximal limb and bulbar weakness, dysphagia, generalized fasciculations with predominance of facial muscles, gynecomastia, and impaired fertility. This study aimed to investigate the correlation of tongue pressure with clinical characteristics and functional measures in patients with KD.

Methods

This study enrolled genetically confirmed KD patients who underwent Videofluoroscopic Swallowing Study (VFSS) and tongue pressure assessment. Patients who had a history of other dysphagia-related diseases such as stroke or head and neck cancer were excluded. VFSS was analyzed by using the Video-fluoroscopic Dysphagia Scale (VDS) and the Penetration Aspiration Scale (PAS). Tongue pressure was evaluated using TPS-100 (CyberMedic, South Korea). Pearson correlation coefficients of maximum tongue pressure with clinical characteristics and functional measures were calculated. Statistical analyses were performed using the statistical software R version 3.6.0.

Results

A total of 30 patients with a mean age of 55.5 ± 9.23 were enrolled. The mean value of maximum tongue pressure (hPa) was 254 ± 86.4 . VDS correlated negatively and significantly with maximum tongue pressure ($r=-0.466$, $p=0.009$). ALSFRS-R and 6-Minute Walk Test correlated positively and significantly with maximum tongue pressure ($r=0.583$, $p<0.001$ and $r=0.375$, $p=0.041$). On the other hand, age, CAG repeat number, and PAS showed no significant correlations with maximum tongue pressure ($r=-0.180$, $p=0.341$; $r=-0.018$, $p=0.925$; $r=-0.116$, $p=0.543$).

Conclusion

Maximum tongue pressure was associated with functional measures including swallowing, physical ability, and gait capacities. This study suggests that tongue pressure may be an indicator of functional status in patients with KD.

Table 1. Clinical characteristics of the patients (n=30)

Characteristics	Value
Age (years)	55.5 ± 9.23
Male	30 (100%)
CAG repeat number	46.3 ± 3.73
Maximum tongue pressure (hPa)	254 ± 86.4
Penetration Aspiration Scale	2.47 ± 1.36
Videofluoroscopic Dysphagia Scale	28.3 ± 10.1
ALSFRS-R	39.2 ± 4.23
6-Minute Walk Test (meters)	302 ± 121

Variables are presented as number (%) or mean ± standard deviation.

ALSFRS-R, Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised.

Table 2. Correlation coefficients of maximum tongue pressure with clinical characteristics and functional measures

Parameter	Pearson's r (95% CI)	p-value
Age	-0.180 (-0.507, 0.193)	0.341
CAG repeat number	-0.018 (-0.376, 0.345)	0.925
Penetration Aspiration Scale	-0.116 (-0.457, 0.255)	0.543
Videofluoroscopic Dysphagia Scale	-0.466 (-0.708, -0.127)	0.009*
ALSFRS-R	0.583 (0.282, 0.780)	<0.001*
6-Minute Walk Test	0.375 (0.017, 0.648)	0.041*

CI, Confidence Interval; ALSFRS-R, Amyotrophic Lateral Sclerosis Functional Rating Scale-

Revised

*p<0.05

Table 3. Scatterplots of various variables. Red boxes and asterisks indicate statistical significance.

