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Tongue pressure during swallowing is decreased in patients with Duchenne muscular dystrophy

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Abstract

Although dysphagia is a life-threatening problem in patients with Duchenne muscular dystrophy (DMD), the pathophysiology of oral stage dysphagia is yet to be understood. The present study investigated the tongue motor deficit during swallowing in patients with DMD and its relationship with disease-specific palatal morphology. Tongue pressure during swallowing water was recorded in 11 male patients with DMD and 11 age- and sex-matched healthy subjects using an intra-oral sensor with five measuring points, and the state of tongue pressure production was compared between the groups. Palatal morphology was assessed by a non-contact three-dimensional scanner on maxillary plaster models. In patients with DMD, the normal sequential order of tongue-palate contact was lost and the maximal magnitude and integrated value of tongue pressure on the mid-anterior part of palate were smaller than those in healthy subjects. The width of the palate in patients was greater than that in healthy subjects and the depth of the palate in patients had a negative correlation with tongue pressure magnitude on the median palate. Our results suggested that the deteriorated tongue motor kinetics prevented tongue movement during swallowing that was appropriate for the depth of the palate and affects the state of tongue pressure production during swallowing.

Keywords: Duchenne muscular dystrophy; Dysphagia; Tongue; Pressure; Swallow

1. Introduction

In Duchenne muscular dystrophy (DMD), dysphagia appears with the progression of the disease [1–4]. There is growing interest in the maintenance of quality of life along with gains in mean life expectancy for patients with DMD and control of dysphagia has attracted attention as an important challenge. However, there have been few reports of dysphagia in patients with DMD. In particular, only a few studies have investigated the oral stage of swallowing, most of which used qualitative observation by videofluorography [2–5].

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The contact pressure between the palate and the tongue, i.e., tongue pressure, makes the greatest contribution to bolus transfer in the oral stage of swallowing [6]. Recently, a tongue pressure measuring system with an ultrathin sensor sheet has demonstrated the normal patterns [7] and age-related changes [8] in tongue pressure production during swallowing in healthy subjects, and a close relationship between abnormalities in the tongue pressure patterns and dysphagia in patients following strokes [9–11].

In DMD, atrophy of muscles and fatty changes in the tongue may cause pseudohypertrophy and macroglossia [12–15] from which increased tongue volume impairs tongue motility [5,16–18]. It was also reported that patients with DMD tend to have wider and shorter hard palates [12,18–22]. Based on these backgrounds, we hypothesized that the morphological and functional abnormalities of the tongue and palate in patients with DMD may cause disease-specific tongue disability during swallowing. Using the tongue pressure measuring system and a non-contact three-dimensional shape measuring system, we quantitatively analyzed the tongue pressure production during swallowing and its correlation with palatal morphology in patients with DMD.

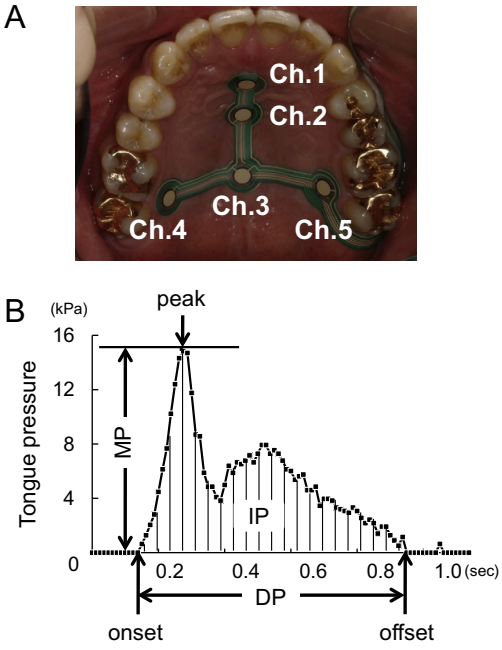

Fig. 1. (A) Tongue pressure sensor sheet. The tongue pressure sensor sheet is attached to the palatal mucosa. Channel (Ch.) 1 was set at the anterior-median part, Ch. 2 at the mid-median part, Ch. 3 at the posterior-median part, Ch. 4 at the right circumferential part, and Ch. 5 at the left circumferential part on the hard palate. To meet these criteria as closely as possible, the appropriate sensor sheet was chosen from three different sizes available (S, M, and L) according to the size of the palate. (B) Items for analyzing the tongue pressure waveforms. MP, maximal magnitude of tongue pressure; DP, duration of tongue pressure; IP, integrated values of tongue pressure.

2. Patients and methods

2.1. Subjects

At the Department of Neurology, National Hospital Organization Toneyama National Hospital, 11 male subjects (age range, 17–24 years; mean age, 20.6 ± 2.3 years) with DMD who were allowed oral feeding without liquid aspiration, could be disengaged from mechanical ventilation for the experiment, could keep stable upright position, and could keep their mouth open widely as determined by their physicians, were enrolled in the DMD group. Details of each subject such as age, diagnosis, Vignos scale, body mass index (BMI), Forced Vital Capacity (FVC), %FVC, ventilation, and the amount of liquid bolus for each subject are summarized in Table 1.

Eight of 11 subjects received a diagnosis from genetic testing by the Multiplex Ligation-dependent Probe Amplification method (MLPA) and 2 of the remaining 3 subjects received a diagnosis from muscle biopsy, which showed a total lack of dystrophin expression despite no abnormality in MLPA. The final subject, who refused to undergo muscle biopsy, received a diagnosis clinically.

Table 1
Details of subjects with Duchenne muscular dystrophy (DMD).

No.	Age	Diagnosis	Vignos scale	BMI (kg/m^2)	FVC (cc)	% FVC (%)	Ventilation	Feeding	FOIS	Bolus volume (ml)
1	18	Gene	9	20.3	810	19.8	NIV part time	Oral	6	10
2	21	Immunohistochemical	9	28.0	1270	31.3	NIV part time	Oral	6	10
3	23	Gene	9	10.5	750	19.2	NIV part time	Oral, tube	3	5
4	23	Clinical	9	9.8	430	10.5	NIV part time	Oral	6	10
5	17	Gene	9	35.2	3190	90.6	None	Oral	6	10
6	18	Gene	9	10.4	640	16.1	NIV part time	Oral	6	10
7	21	Gene	9	12.9	–	–	NIV full time	Oral	5	10
8	22	Gene	9	18.9	–	–	NIV full time	Oral	5	10
9	24	Gene	9	15.6	–	–	NIV full time	Oral	4	5
10	21	Immunohistochemical	9	11.0	590	13.9	NIV part time	Oral	5	5
11	20	Gene	9	21.6	400	10.2	NIV part time	Oral	6	10

BMI: body mass index; FVC: forced vital capacity; FOIS: functional oral intake scale; NIV: non-invasive ventilation.
In subject 7–9, respiratory function tests were not performed because of using ventilator the whole day.