

# Longitudinal Changes of Tongue Thickness and Tongue Pressure in Neuromuscular Disorders

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## Research article

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# Abstract

**Background:** Swallowing dysfunction is related to major cause of adverse events and an indicator of shorter survival among patients with neuromuscular disorders (NMD). It is critical to assess the swallowing function during disease progression, however, there are limited tools that can easily evaluate swallowing function without using videofluoroscopic or videoendoscopic examination. Here, we evaluated the longitudinal changes in tongue thickness (TT) and maximum tongue pressure (MTP) among patients with amyotrophic lateral sclerosis (ALS), myotonic dystrophy type 1 (DM1), and Duchenne muscular dystrophy (DMD).

**Methods:** Between 2010 and 2020, TT and MTP were measured from 21 ALS, 30 DM1, and 14 DMD patients (mean ages of 66.9, 44.5, and 21.4 years, respectively) at intervals of more than half a year. TT was measured, by ultrasonography, as the distance from the mylohyoid muscle raphe to the tongue dorsum, and MTP was determined by measuring the maximum compression on a small balloon when pressing the tongue against the palate. Then we examined the relationship between these evaluations and patient background and swallowing function.

**Results:** Mean follow-up periods were 24.0 months in the ALS group, 47.2 months in the DM1 group, and 61.1 months in the group. The DMD group demonstrated larger initial TT than the other groups, while the DM1 group had lower initial MTP than the ALS group. The ALS group showed a greater average monthly reduction in mean TT than the DM1 group and greater monthly reductions in mean BW and MTP than the other groups. Significant differences between the first and last BW, TT, and MTP measures were found only in the ALS group.

**Conclusions:** This study suggests that ALS is associated with more rapid degeneration of tongue function over several years compared to DMD and DM1.

## Background

Patients with neuromuscular disorders (NMDs) often experience problems with swallowing during the course of the illness. They require a periodic video fluoroscopic swallowing study (VFSS) or fiberoptic endoscopic evaluation of swallowing (FEES) to reduce the risk of aspiration pneumonia and assess the need for appropriate nutritional support. However, these tests require equipment and can only be evaluated in specialized facilities, and it is difficult to assess tongue function, using VFSS or FEES, quantitatively. Clinically, tongue function is determined by multiple characteristics, including mobility, shape, and posture. In NMDs, dysphagia may result from tongue muscle weakness as well morphological changes due to muscle hypertrophy or atrophy. For instance, an enlarged tongue is frequently observed in Duchenne muscular dystrophy (DMD) while tongue atrophy is common in amyotrophic lateral sclerosis (ALS), both of which may cause dysphagia. Therefore, monitoring these changes is critical for clinical management during disease progression.

In our previous study, we evaluated the relationship between tongue thickness (TT) and strength in NMD patients at a single time point [1] and found that TT was significantly greater in DMD patients, while maximum tongue pressure (MTP) was lower in myotonic dystrophy type 1 (DM1) patients compared to other NMD groups. Moreover, that study revealed a significant correlation between TT and MTP in ALS. These findings indicate that association between TT, MTP, and dysphagia varies depending on the type of NMD. However, longitudinal changes of TT and MTP during the course of NMD have yet to be studied. This information is critical for evaluating aspiration and malnutrition risk over time. This current study describes the longitudinal changes in TT and MTP as well as body weight (BW) to help establish disorder-specific treatment plans for dysphagia in DMD, DM1, and ALS patients.

## Methods

### Subjects

Between 2010 and 2020, TT and MTS were measured from 21 ALS patients (9 males and 12 females; mean age, 66.9 years), 30 DM1 patients (16 males and 14 females; mean age, 44.5 years), and 14 male DMD patients (mean age, 21.4 years) at the Department of Neurology, Neuro-Muscular Center, NHO Omuta National Hospital, Omuta City, Japan. Patients received multiple TT and TP measurements at intervals of more than half a year. All DMD and DM1 cases were confirmed by genetic analysis after clinical diagnosis, and all ALS patients were clinically diagnosed as probable or definite by neurologists specializing in ALS, based on Awaji criteria [2]. Furthermore, all ALS patients were monitored clinically for more than one year before enrolling in this study. These 21 patients were divided into 14 bulbar onset and 7 limb onset subgroups for analysis. TT and MTP measurements and VFSS are conducted as part of standard examinations in the NHO Omuta National Hospital and the ethics committee (or institutional review board) of the NHO Omuta National Hospital approved the analysis of the examinations in this study. Informed consent was obtained through an opt-out provision on the study website. Physical examinations were conducted on all patients, including measures of BW, height, and body mass index (BMI).

### Tongue thickness

TT was measured from the mylohyoid muscle raphe to the upper surface of the tongue (Fig. 1) using a Fukuda Denshi UF-550XTD ultrasound system (Fukuda Denshi, Tokyo, Japan). The transducer and equipment settings were monitored constantly to obtain optimal ultrasound images for quantitative analysis [3]. Measurements were performed three times for the submental muscle group, and the average value was calculated. The TT was measured in a sitting position with relaxed normal head and neck posture and relaxed closed mouth. Excessive pressure on the skin during scanning was avoided by the generous use of contact gel.

### Measurement of Tongue Strength

Tongue strength was measured as MTP using a handy probe consisting of a small balloon pressurized with air to 19.6 kPa (JM-TPM; JMS, Tokyo, Japan) [4]. Each participant was required to compress the balloon against the palate using the tongue for approximately 7 s while applying maximum effort. The resulting increase in balloon inner pressure was measured and recorded as MTP. This test was repeated three times, and the mean value was obtained for analysis.

## Data Analysis

Mean BW, TT, and MTP were compared among groups using one-way ANOVA with Bonferroni multiple comparisons post-test. Pearson correlation coefficients were calculated to assess the relationships among BW, TT, and MTP. The first and last BW, MTP, and TT measurements were compared by the paired *t*-tests. All statistical analyses were performed using SPSS 13.0 J for Windows (SPSS, Inc., Chicago, IL, USA). A  $p < 0.05$  (two tailed) was considered statistically significant for all tests.

## Results

Patients clinical characteristics are summarized in Table 1. As expected, mean age was higher, while follow-up duration was significantly shorter in the ALS group. Initial mean BW was significantly lower in the DMD group than the DM1 group ( $p < 0.01$ ). The mean initial TT was larger in the DMD group than the other groups ( $p < 0.01$ ), while MTP was lower in the DM1 group than the ALS group ( $p < 0.05$ ).

However, the ALS group demonstrated the greater average monthly reduction in mean BW than the other two groups ( $p < 0.01$ ), the greater average monthly reduction in mean TT compared to the DM1 group ( $p < 0.05$ ), and the greater average monthly reduction in mean MTP compared to both the DM1 and DMD groups ( $p < 0.01$ ). Moreover, only the ALS group showed significant differences in BW, TT and TP between the initial and final measurements ( $p < 0.01$ ,  $p < 0.05$ ,  $p < 0.01$ , respectively) (Fig. 2a, b).

Analysis of individual subjects revealed TT and MTP were reduced in 15 of 21 ALS patients (Fig. 3a, b). Subgroup analysis revealed no significant difference in the incidence of reduced TT between bulbar and limb onset patients, while none of the ALS patients with bulbar onset had a substantial reduction in MTP. There were no significant correlations among BW, MTP, and TT reduction rates of BW, MTP, and TT in any NMD group.

## Discussion

In our previous cross-sectional study, we evaluated the relationship between TT and MTP as a strength metric in DMD, DM1, and ALS patients. There was higher TT in the DMD group, lower MTP in the DM1 group, and a significant correlation between TT and MTP in the ALS group [1]. In another study, we also revealed the distinctive features of dysphagia in DM1 and DMD patients using VFSS, hyoid bone movement during the pharyngeal phase of swallowing (excursion), or pharyngeal residue measurement [5], which also demonstrated distinct abnormalities in swallowing muscle function between DM1 and DMD. The study also showed the correlation between TT and BW, but not MTP, in the DM1 group [1],

suggesting that, in DM1 patients, TT was associated with malnutrition and concomitant weight loss. However, long-term tongue muscle weakness is also thought to promote malnutrition, and further loss of muscle mass may lead to additional TT and BW reductions [6]. Therefore, we assessed TT, MTP, and BW at multiple times during disease progression in this study.

Van Den Engel-Hoek et al. reported that TT can be assessed, conveniently and reproducibly, in DMD patients using ultrasound [3, 7]. More recently, they reported increased tongue hypertrophy and dystrophic changes in masticatory muscles but did not report changes in tongue muscles [8]. Although we found “tongue pseudohypertrophy” in DMD patients by ultrasound assessment, there were no significant longitudinal changes as in the ALS group. We suggest that, once tongue muscle tissue is replaced by connective tissue or fat in the early stage of DMD [9], there is no further progression in term of TT.

Poor tongue strength is independently associated with shorter survival time in ALS patients [10], so measuring TT and MTS is critical for prognosis and adjustment of treatment. Tamburrini et al. examined tongue movement function among ALS patients using both ultrasonography and VFSS, but they did not conduct objective tongue atrophy assessment [11]. Nakamori et al. suggested an association between TT, disease progression, and tongue dysfunction [12], consistent with our previous study showing a significant correlation between TT and MTP in ALS [1]. The mean TT value of ALS patients in their study ( $41.9 \pm 4.0$  mm) was similar to our final measurement ( $40.0 \pm 6.5$  mm), while the mean TT value of their control volunteers ( $44.8 \pm 3.0$  mm) was closer to the initial value in our ALS group ( $42.8 \pm 6.8$  mm), strongly suggesting tongue atrophy progressed during the roughly 2-year follow-up period. Nakamori et al. also found reduced TT over 15 months. In accordance with their study, the ALS group also included patients with no clear TT reduction (Fig. 3a), possibly due to differences in time from onset or rate of disease progression between bulbar or limb onset types. Alternatively, we found no association between TT and BW, in contrast to the linear association between TT and BMI reported by Nakamori et al. In our previous study, bulbar onset patients showed a stronger correlation between TT and MTP than limb onset patients, suggesting these measures especially important for monitoring bulbar paralysis patients [1], especially during the early clinical stages.

This study has several limitations, most notably the differences in mean age among groups, which is known to influence MTP [13]. However, age matching of DMD, ALS, and DM1 is difficult due to the differences in age at onset and course. Nonetheless, these results suggest distinct dysphagia pathomechanisms and progression features of among neurogenic and myogenic disorder patients.

This longitudinal study revealed more rapid loss of tongue thickness and strength in ALS patients than DMD and DM1. Only the ALS patients also exhibited substantial weight loss over the observation period, a change strongly associated with shorter survival [14]. On the other hand, DM1 and DMD patients may show only slowly developing tongue dysfunction in the short term, although DM1 may have lower tongue strength and DMD patients enlarged tongues, respectively, both of which can contribute to dysphagia. These findings suggest that regular re-evaluation of TT and MTP could provide valuable information on tongue dysfunction progression in NMD.

## Conclusions

For assessment of tongue function, TT and MTP measurements are more convenient than VFSS and FEES. This study suggests that the relationships between TT, MTP, and swallowing function differ depending on NMDs and ALS is especially associated with more rapid degeneration of tongue function over several years compared to DMD and DM1.

## Abbreviations

NMDs, neuromuscular disorders; VFSS, video fluoroscopic swallowing study; FEES, fiberoptic endoscopic evaluation of swallowing; DMD, Duchenne muscular dystrophy; ALS, amyotrophic lateral sclerosis; TT, tongue thickness; MTP, maximum tongue pressure; DM1, myotonic dystrophy type 1; BM, body weight; BMI, body mass index

## Declarations

### *Ethics approval and consent to participate*

TT and MTP measurements and VFSS are conducted as part of standard examinations in the NHO Omuta National Hospital and the ethics committee (or institutional review board) of the NHO Omuta National Hospital approved the analysis of the examinations in this study. The committee's reference number is 2-20. Informed consent was obtained through an opt-out provision on the study website.

### *Consent for publication*

Not applicable

### *Competing interests*

The authors declare that they have no competing interests" in this section.

### *Availability of data and materials*

Most of the data can be obtained from the corresponding author if necessary.

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### *Authors' contributions*

GU was involved in the research project, the statistical analysis, and writing the manuscript. SF, HF and YT were involved in the review and critique of the manuscript. HA, NoS, MT, and NaS were involved in the organization and execution of the research project.

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## Tables

Due to technical limitations, table 1 is only available as a download in the Supplemental Files section.

## Supplementary Files

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