Time dependent changes of variables associated with malocclusion in patients with Duchenne muscular dystrophy

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Time dependent changes of parameters associated with malocclusion in patients with Duchenne muscular dystrophy (DMD) were examined in four dental developmental stages in 34 patients. We adopted activities of daily living (ADL) score, dental arch, craniofacial morphology, and electromyograms of the masseter and temporalis muscle as parameters. A comparison was made with the results in DMD subjects to data from healthy subjects with normal occlusion reported in the literature. In DMD subjects, manifestations of open-bite were related to ADL score, sagittal shortening and transverse expansion of the dental arch and vertical overgrowth of the lower jaw. Posterior cross-bite malocclusion was associated with differences in the time dependent changes between the jaws in transverse expansion. The malocclusion in DMD subjects was also related to the time dependent disproportional changes in masticatory muscle function by EMG. Occlusal deviation in DMD subjects became apparent at the late mixed dentition and malocclusion became definitely manifest from early permanent dentition.

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INTRODUCTION

The Duchenne type of progressive muscular dystrophy (DMD) is a group of genetically and clinicaly different myopathies characterized by progressive atrophy of the skeletal muscle and decreased muscle strength. DMD shows sex-linked recessive inheritance and occurs mainly in males. In most cases, gait disturbance and falling due to decreased psoas muscle strength become apparent between 2 and 5 years of age. As symptoms gradually progress, ambulation usually is lost at 12 to 13 years. Skeletal deformity occurs as a result of muscle contractures. Respiratory failure due to muscle atrophy develops in the terminal stage. Most patients die during the second decade of life or the beginning of the third decade often from respiratory complications. DMD is described as psuedohypertrophy in muscle biopsy specimens. Marked fatty infiltration can be identified as the cause of psuedohypertrophy. The masseter and temporalis muscles are among those showing such psuedohypertrophy and for similar reasons macroglossia also occurs.

Dental investigators have noted abnormalities in the dental arch configuration and occlusal relationships as a result of the widespread muscle changes in DMD and also because of a decrease in masticatory function. In particular, open-bite (anterior and extending to posterior) is a common finding in patients with DMD. This change is progressive and associated with systemic and local alterations.[6,8,10,11] The relationships among factors causing malocclusion in DMD have not yet been clarified enough.

Recently, Eckhardt and Harzer reported that circumoral muscle imbalance is closely related to malocclusion in DMD patients.[8] Kikiaridis and Katsaros[10] reported in his review regarding the influence in dentofacial morphology of these diseases on the orofacial muscles that the posterior crossbite in DMD is due to the transversal expansion of the mandibular arch, possibly because of the decreased tonus of the masseter muscle near the molars in combination with the enlarged hypotonic tongue and the predominance of the less affected orbicular oris muscle. However, there were few studies which comprehensively evaluated the relationship of changes in dental arch form and occlusal pattern with masticatory muscle function and age-related progression of DMD.

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This study was presented in part at the 14th Congress of the International Association of Dentistry for the Handicapped and the 15th Annual Meeting of the Japanese Society of Dentistry for the Handicapped in September 1998, Yokohama, Japan.

We examined causal factors for malocclusion in patients with DMD by correlating malocclusion with changes in activities of daily living (ADL) score, dental arch, craniofacial morphology, and electromyograms of the masseter and temporalis muscle between the two groups. In addition, we examined time dependent changes of malocclusion in the subjects in comparison with healthy children.

SUBJECTS, MATERIALS AND METHODS

Subjects

All protocols involving human subjects were approved by the Institutional Review Committee of Kagoshima University. The study was performed in accordance with the Helsinki Declaration of 1975 as revised in 1983.

The patients consisted of 34 males with DMD diagnosed by neurologists. They were grouped two from early mixed dentition to early permanent dentition (1-G) and full permanent dentition (II-G) (Table 1). Mean patient age was 15.0 years. Data obtained included diagnostic dental casts, lateral roentogenocephalographs, electromyographs (EMG) and activities of daily living (ADL) scores. The number of subjects varied in different data analysis because some types of data could not be obtained in certain patients (Table 1).

For control data we used previous reports of observations in healthy subjects.[12,13] These subjects (56 for morphology and 21 for EMG) had normal occlusion, no cranio-facial skeletal abnormality and no abnormality of dentition. Gender distribution and mean and for these subjects are shown in Table 2. They will be referred to as "control data" through out this report.

Table 1. DMD patients by group and used data.

Group Number of Subjects (Mean Age) by group	I-G 17(11.0)	II-G 17(17.2)	Total 34(15.0)
Activities of Daily Living (ADL)	17	14	31
Lateral roentgenocephalogram	17	13	30
Dentition	17	16	33
EMG	16	10	26

Table 2. Healthy children with normal occlusion for control data.

Group	Dental stage	Healthy children in morphology				Healthy children in EMG			
		male	female	total	mean age	male	female	total	mean age
	III-A	6	9	15	8.5	3	3	6	8.0
I-G	III-B	6	9	15	10.6	2	2	4	10.6
	III-C	5	6	11	12.4	3	2	5	12.1
II-G	IV-A	6	9	15	15.0	3	3	6	15.1
	total	23	33	56	11.6	11	10	21	11.5

By Iwasaki *et al* [12,13]

Activities of daily living (ADL) score

Using Ueda's method,[14] 25 items related to activities of daily living of importance to Japanese DMD patients were rated using a 0 - 4 scoring system. A perfect score was 100 or 100%. and subtracting from 100 when an item score was <4 performed scoring. The 25 items are listed in Table 3.

Measurements of the dental and study casts. As shown in Figure 1, length and width in a dental arch cast were measured using an automatic digital caliper (NTD12-15®, Mitutoyo. Tokyo. Japan).[15,16] Anterior open-bite was defined as a negative over bite between the maxillary and mandibular right central incisors.

Table 3. Items of activities of daily living.

1. Walking,	13. Seating,
2. Standing,	14. Raising one's both hands,
3. Going up the stairs,	15. Putting on one's undershirt,
4. Going down the stairs,	16. Putting one one's trousers,
5. Standing up from the floor,	17. Fix and not sway the neck,
6. Standing up from the chair,	18. Laying oneself on the bed,
7. Sitting down on the chair,	19. Turning over in the bed,
8. Keep on talking with breath,	20. Rising,
	21. Crawling,
9. Writing,	22. Using the toilet.,
10. Washing one's face,	23. Taking a bath,
11. Wiping with a towel,	24. Holding a bowl at a meal,
12. Squatting down,	25. Riding a wheelchair,

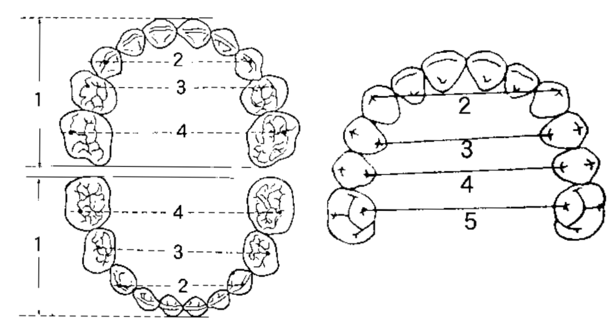

Figure 1. Landmarks and reference lines of the primary dental arch (left) and permanent (right).

1. length of the dental arch:
2. width of the canines:
3. width of first premolars or first deciduous molars:
4. width of second premolars or second deciduous molars:
5. width of first permanent molars

Lateral roentgenocephalograms

We measured eight skeletal angles, four dental angles, and eight lines. Measured items are illustrated in Figure 2.

EMG recording

Six channels were processed simultaneously in a polygraph system (RM-6000®: Nihon Kohden, Tokyo,

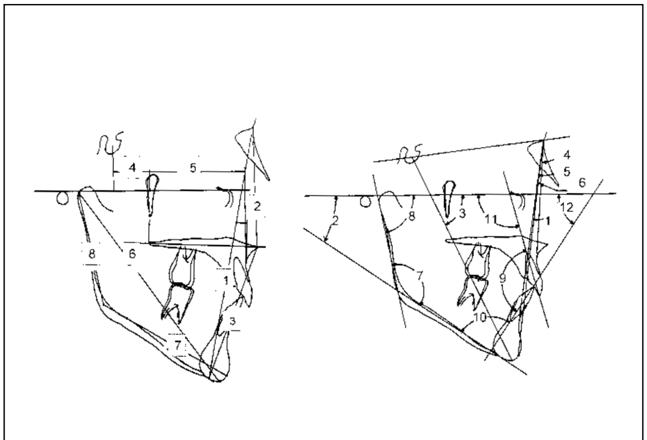

Figure 2. Linear (left), skeletal and dental angular (right) measurements in lateral roentgenocephalogram.

Linear measurements 1. N-Me, 2. N-ANS, 3. ANS-Me, 4. S-Ptm, 5. A-Ptm, 6. Gn-Cd, 7. Pog-Go, 8. Cd-Go

Skeletal and dental angular measurements 1. Facial Angle, 2. Mand.P A., 3. Y-axis, 4. SNA, 5. SNB, 6. ANB, 7. Gonial Angle, 8. ramus inclination, 9. Interincisal Angle, 10. L 1 to Mandibular Plane 11. U1 toFH., 12. FMIA

Japan) and recorded using a Thermal Array Recorder (RTA-1300®; Nihon Kohden}. The time constant was 0.01 sec and a 10-mm deflection was calibrated to correspond to 200mV of input. Paper speed was 25mm sec. The simultaneously evaluated muscle sites were the anterior temporalis (TA) and the posterior portion (TP) of the same muscle, as well as the superficial portion of the massetermuscle (M) on both right and left. Concentric bipolar miniature skin electrodes (Ag/Cl gel 5 mm internal diameter and 12 mm external diameter: Nihon Kohden) were affixed to the skin surface using electrode paste and tape at points overlying each muscle site tested. Electrodes were kept at least 2.0 cm apart. The ground was attached to the right earlobe. After the posture was adjusted, they were asked to chew gum (Lotte, Tokyo, Japan) at a rate of about 76 times/min,[12] and also were asked to clench their teeth. In addition to thermal paper recording, muscle potential waveforms were recorded using a universal data recorder (A-613: Sony, Tokyo, Japan) and data were processed as described below.

Processing of EMG data

Integration

An "if-800 data processor" (model 1150, OKI Denki, Tokyo, Japan) with a central processing unit (12-bit, 16-channel multiplexed mode) and an analog-digital (AD) convener with 40 kHz conversion capacity was used. AD conversion for this study involved six channels at 1 kHz. Recorded analog data were convened to digital format at 1 mm see by the AD convener: 4096 samples covering about 4 sec were quantified and the six channels were stored simultaneously using a disk cartridge.

Conversion voltage ranged from -5.0 to + 5.0V and -5.0V was converted to -2048 and +5.0V to +2047. The tape speed was 9.5cm see during electromyographic recording and 2.4cm see during sampling. After a calibration pulse was input and stored as a standard for conversion of data to microvolts. Electromyograms for each type of action were input and integrated using Sympson's primary numerical integral method. On electromyograms obtained during gum chewing to evaluate rhythm. The first five and last five strokes were excluded with the intervening five strokes being evaluated.

In maximal clenching for evaluating integral value, five waves were obtained and the maximal three consecutive waves representing the middle 1.0sec of each jaw clenching were studied.

Calculations were performed to convert the integral value to microvolts per second. The integral values for TA, TP, and M on the right and left sides in five strokes during gum chewing were added.

The distribution percentage of each muscle examined was calculated using this total action potential as the denominator. The value obtained was defined as a relative interval value ratio (RIVR) and used as final data for analysis.

Rhythm of masseter muscle by EMG

Rhythm of the EMG in M was measured using an automatic digital caliper (Mitutoyo, Tokyo, Japan) that was accurate to 0.01 mm. Measurements were made in 20 early strokes on electromograms during gum chewing obtained using an ink-writing oscillograph. Intervals in the tracing were converted to seconds. Each stroke was assessed in terms of three intervals as follows:[12] Duration (DR) was the duration of discharge: the interval (IT) was the time between two discharges: D+I was defined as a cycle: and D/C was defined as a DR/D+I. The mean value and coefficient of variation were determined for each interval of M. Similarly, the mean value and coefficient of variation for each interval was obtained for each 5 of the 20 strokes.

STATISTICS

After confirming data reproducibility, one sample t test was performed to examine the differences between the DMD subjects and control data in each measurement. The relationship between the two groups was assessed statistically by two samples t test with Welch's correction using the value of differences between the DMD subjects and control data in each measurement.

Statistical significance was accepted when the probability (p) was less than 0.05.

RESULTS

Prevalence of anterior open bite by group

The number of DMD subjects with anterior open-bite was markedly high in II-G (76.5%) comparing with l-G (35.3%). Average open bite was -2.0mm in I-G and -5.6mm in II-G (Table 4).

Table 4. Status of anterior open-bite in DNID patients.

Group	I-G	II-G	Total
Total subjects	17	17	34
Anterior open-bite subjects	6	13	16
Frequency of anterior open-bite (%)	35.3	76.5	-
Average over bite (mm)	-2.0	-5.6	-

ADL score

ADL score decreased significantly (p<0.001) with group in all DMD subjects. In II-G, ADL scores in the DMD with anterior open-bite occlusion were significantly lower (p< 0.01) than in the DMD without anterior open-bite (Figure 3).

Dental arch cast measurement

Figures 4 and 5 show differences in dental arch measurements between groups (DMD subjects and control data).

Maxillary dentition

Data from DMD subjects did not differ significantly between groups or from control data in maxillary length. Both the first permanent premolars or first deciduous molars distance [4-4 (D-D)] and the second permanent premolars or second deciduous molars distance [5-5 (FE)] were significantly more narrow than control data in l-G (p<0.01), but the first permanent molars distance (6-6) was significantly wider (p<0.05). However, the canines [3-3 (C-C)], 4-4 (D-D), 5-5(E-E)], and 6-6 were significantly more wide than control data in II-G (p<0.001, p<0.001).

In comparison the two groups, l-G with II-G, the 4-4 (D-D), the 5-5 (F-F), and the 6-6 in II-G were significantly wider than in I-G (p<0.001, p<0.001, p<0.001).

Mandibular dentition

When we compared the length of the lower dental arch in DMD subjects and control data, we found that they were similar even though the length in the DMD was significantly shorter than in control data (p<0.001) in total. In addition, mandibular length was significantly shorter (p<0.05) than control data in I-G. When we compared the widths, 3-3 (C-C), 4-4 (D-D), 5-5 (E-E) and 6-6 in each group, 3-3 (C-C) and 6-6 were significantly wider than control data in I-G (p<0.05, p<0.0001). The other side, each width in II-G [the 3-3 (C-C) the 4-4 (D-D), the 5-5 (F-F), and the 6-6] was significantly wider than control data (p<0.0001).

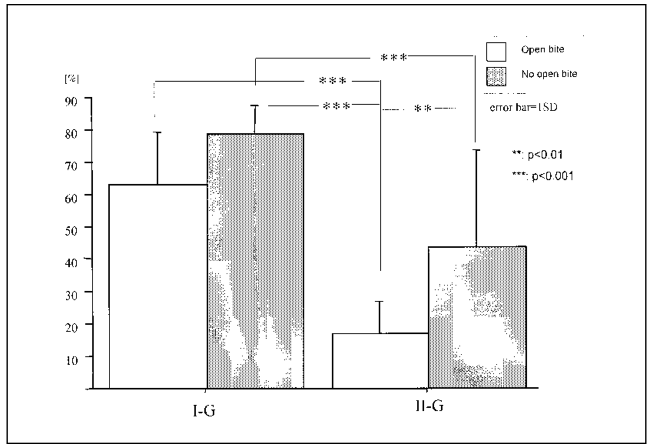

Figure 3. ADL score by group comparing status of anterior bite.

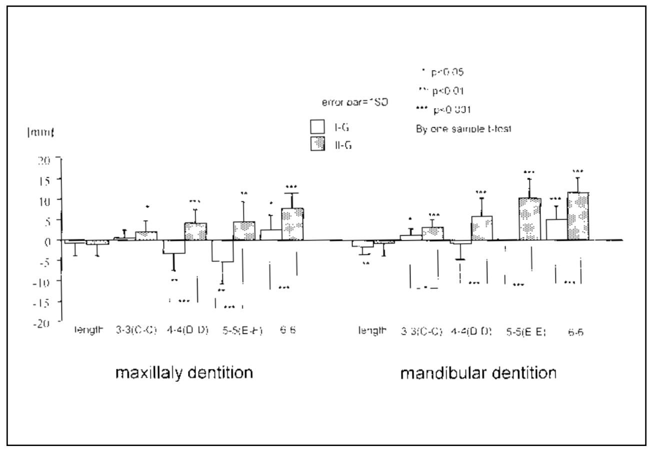

Figure 4. Difference between DMD and control by length and width comparing groups. The same data in DMD to control is zero.

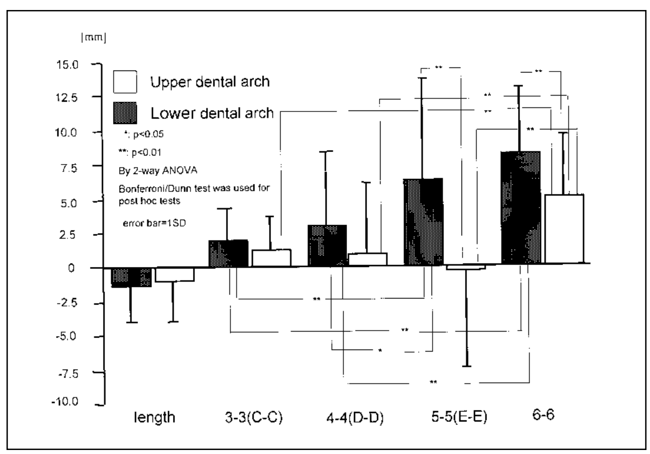

Figure 5. Difference between DMD and control for each measurement of length and width of dental arch.

In comparison, the two groups I-G with II-G each width in II-G [the 3-3 (C-C), the 4-4 (D-D), the 5-5 (F-F), and the 6-6] was significantly wider than in I-G (p<0.05, p<0.001, p<0.001, p<0.001). Posterior measurement sites showed greater differences between the DMD and con-

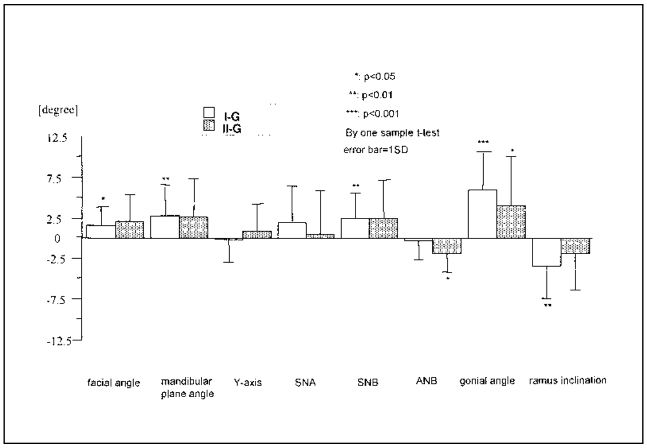

Figure 6. Difference between DMD and control for 8 skeletal pattern angles in lateral roentgenocephalograms, comparing groups.

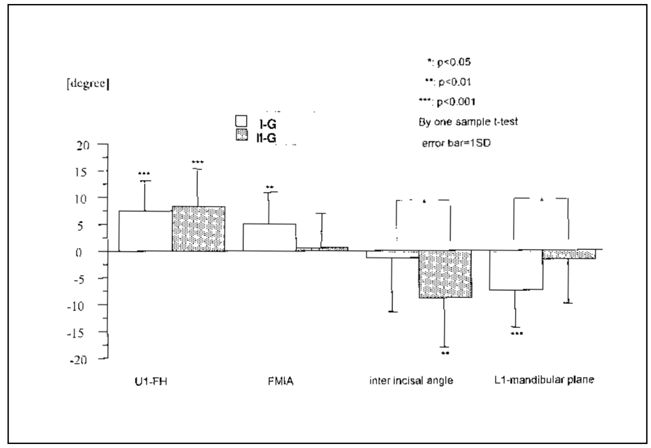

Figure 7. Difference between DMD and control for 4 dental pattern angles in lateral roentgenocephalograms, comparing groups.

trol data than anterior measurement sites. The difference between the DMD and control data for 6-6 in both dental arches was significantly larger (p<0.01) than the difference for 3-3 (C-C) or 4-4 (D-D).

Differences for 5-5 (F-F) and for 6-6 between the DMD and control data were greater for mandibular dentition (p<0.01) than for corresponding comparisons in maxillary dentition.

In comparison both jaws, lower with upper, the 6-6 of mandibular dentition in I-G was significant wider (p<0.05) than of maxillary dentition. However, both the 5-5 (E-E) and the 6-6 of mandibular dentition in II-G were significantly wider (p<0.01, p<0.01) than the maxillary dentition.

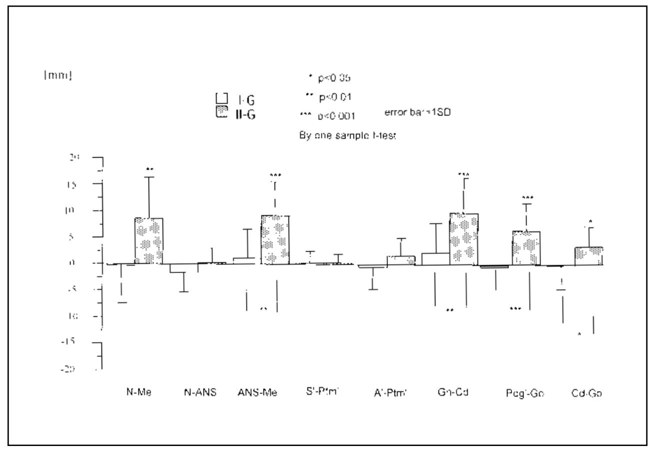

Figure 8. Difference between DMD and control for 8 linear measure items in lateral roentgenocephalograms, comparing groups.

Lateral roentgenocephalograms

Angles

Measured angles in I-G showed significant differences between DMD subjects and control data except for the interincisor angle, Y-axis, SNA, and ANB (Figures 6 and 7). Upper incisor (U1)-to- Frankfort plane angle, Frankfort mandibular incisor angle (FMIA), the facial angle, the mandibular plane angle SNB and the gonial angle were significantly greater than in the control data (p<0.001, p<0.001,p<0.05, p<0.01, p<0.01, p<0.001 respectively). However, the lower incisor (L-1)-to-mandibular plane angle and the ramus inclination were significantly smaller than in the control (p<0.001, p<0.01. The other side, measured angles in II-G showed no significant differences between DMD subjects and control data except for Upper incisor (U1)-to-Frankfort plane angle, the interincisor angle, ANB and the gonial angle. The interincisal angle and ANB were significantly lower than in the control data p<0.05. Upper incisor (U 1)-to- Frankfort plane angle and the gonial angle were significantly greater than in the control data p<0.05.

There were no significant differences between I-G and II-G except for the interincisal angle and the lower incisor (L-1)-to-mandibular plane angle. Both the interincisal angle and the lower incisor (L-1)-to-mandibular plane angle in II-G were significantly smaller than in I-G (p<0.05).

Linear measurements

The linear distances of N-Me (p<0.01), ANS-Me (p<0.001), Gn-Cd (p<0.001), Pog-Go (p<0.001), and Cd-Go (p<0.05), in II-G were significantly longer than in control data (Figure 8). The linear distances of ANS-Me (p<0.01), Gn-Cd (p<0.005), Pog-Go (p<0.001), and Cd-Go (p<0.05), in II-G were significantly longer than in I-G.

EMG of the masticatory muscles

Relative integral value ratio (RIVR)

RIVR of M in I-G was found to be a greater than TA in control data. However, no reversal of RIVR of TA and M in DMD subjects was observed in I-Ct (Figure 9). In I-G RIVA of M in DMD subjects was significantly smaller than in control data (p<0.05).

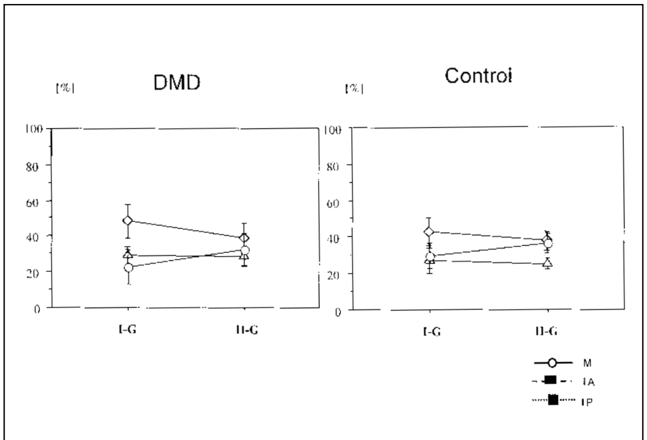

Figure 9. Change of relative integral value ratio on EMG in masseter and temporal muscle by groups.

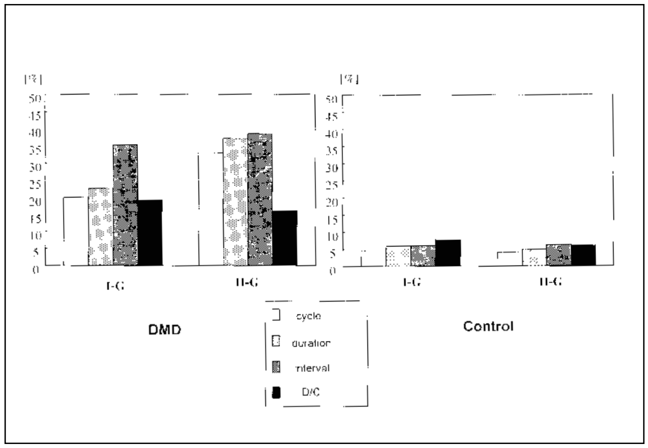

Figure 10. Coefficient of variation of masticatory rhythm on EMG in masseter muscle at each dental stage.

Rhythm in masseter muscle

DR, IT, and D+I in the DMD were significantly longer than in control data for both sides of M, TA and TP in total at least ($p<0.05$).

Concerning stability of rhythm of M, coefficients of variation in the DMD were significantly higher ($p<0.05$), for DR, IT, and D+I. and D/C than in control data in both groups. In particular, the highest coefficients of variation of each rhythm except for D/C in II-G were observed in DMD subjects (Figure 10).

DISCUSSION

Time dependent changes of multiplex findings associated with occlusal deviation in DMD subjects in this study were obtained. It was difficult to discuss clearly in the totality of these findings. Therefore, we discussed the findings as classified in five categories as follows: open bite and other clinical malocclusions in DMD subjects, ADL score, dental arch configuration, measurements in lateral roentgenocephalograms and electromyographs of the masticatory muscles.

Open bite and other clinical malocclusions in DMD subjects

The reported prevalence of anterior open-bite occlusion in DMD subjects is 41.2 to 66%.[17-20] In the present study, the frequency of anterior open-bite, which in some cases was associated with posterior open-bite, was 35.3 in I-G and 76.4% in II-G. The prevalence and degree of anterior open-bite increased with age as in previous studies.[20] More generally, the prevalence of malocclusion, including anterior open-bite occlusion, was reported as 86% in DMD patients.[6,11] Anterior open-bite occlusion occurs much more frequently in DMD patients than in the general population for which the prevalence has been reported as 5.3% in Japan.[21] Many cases of malocclusion in DMD patients combined openbite (anterior and extending to posterior) with cross-bite occlusion.[6,10,11,20] Similarly, combined malocclusion including cross-bite was present in 20% of our DMD subjects.

Higher occurrences have been claimed by Eckardt and Harzer, who reported that they invariably observed a posterior cross-bite in DMD patients.[8] The review reported by Kikiaridis and Katsaros[10] also claimed the relationships among posterior cross-bite, the transversal expansion of the mandibular arch, and the decreased tonus of the masseter muscle near the molars. Several earlier reports have demonstrated anterior-posterior and buccal-lingual asymmetry changes affecting the maxillary and mandibular dental arches to be causal factors for cross-bite occlusion.[17,18,22] In addition, a previous study of DMD patients concluded that transverse expansion of the dental arch could be due to enlargement of a hypotonic tongue.[10] These studies also demonstrated that differences in transverse expansion between the lower and upper jaw reflected differences in timing of pathological functional deterioration between the orbicularis oris and masticarory muscles.[8,10]

Our results and findings of some previous studies suggested that anterior open-bite occlusion and combined malocclusion in DMD patients result from disharmony between three muscle group forces: internal force from inside the dental arch, external force from the buccal or labial aspects, and vertical force for occlusion.

ADL score

ADL score in the DMD subjects decreased with age and showed a marked decrease from late mixed dentition in I-G supporting a previously reported marked decrease in ADL score from age 10 to 13.[20] In our study, ADL scores in DMD subjects with anterior open-bite occlusion were significantly lower than in patients without the open-bite occlusion, in II-G. These findings demonstrate that the development of open-bite is associated with progression of systemic muscular dysfunction.

Dental arch configuration

In DMD patients, the length of the mandibular dental arch has been reported as showing shortening in the sagittal.[8,23] In our study, however, the length of the dental arch in DMD subjects was found to be slightly shorter in only lower jaw with significant in I-G than in control data. In contrast, the lower jaw in DMD subjects was significantly wider ($p<0.0001$-0.05) than in control data in II-G at least. The increase in width first became notable posteriorly and advanced anteriorly with increasing age. This increase occurred earlier and was more expansive in the lower jaw. Our results were consistent with those previous investigations of development of distorted dental arch configuration in DMD patients.[6,8,10,11] Shortened length and increased width of the dental arch have been hypothesized to result from functional imbalance between craniofacial muscle groups.[8,10]

Another study also found the lingual pressure to be higher than bucco-labial pressure even in the normal subjects,[24] lingual pressure upon the teeth is decreases less than buccal or labial pressure because the tongue compensates for declining swallowing function in DMD patients.[25] Even at rest, the tongue may press against the mandibular dentition resulting in widening of the arch. Widening of the arch is related to malocclusion in DMD patients.

In contrast, changes in the posterior portion of the dental arch in DMD subjects were relatively more prominent in earlier age. This finding may reflect the abnormal enlargement of the tongue, progressing over time from the root to the apex. This progression from posterior to anterior tongue is typical for DMD patients. Marked transverse expansion of the dental arch in the posterior area appears to result in posterior cross-bite in DMD patients.

Measurements in lateral roentgenocephalograms

As for skeletal pattern descriptions of skeletal changes in DMD patients have included many reports of gonial angle high values for the mandibular plane angle, Y-axis, gonial angle and SNB, and low position of the lower jaw.[17,18,20,26] These abnormalities are considered to be major factors favoring development of open-bite occlusion in DMD patients.

In our study, the DMD subjects showed significantly higher values than control data for the gonial angle and linear measurement items related to vertical growth of the lower jaw in both groups. These results suggest that open-bite in DMD subjects is associated not only with disproportional changes of the dental arch, but also with disproportion in vertical growth of the lower jaw.

As for dental pattern, high values were reported for Ul to SN in DMD.[18,26,28] In the present study, the DMD subjects showed significantly greater values for upper incisor (U1)-to-Frankfort plane angle in both groups than in control data. However, both the interincisal angle and the lower incisor (L-1)-to-mandibular plane angle in Il-G were significantly smaller than in I-G. These results were consistent with previous reports that open-bite occlusion in the DMD patients was related to the low position of teeth, protrusion of upper incisors, and retrusion of lower incisors.

Electromyography of the masticatory muscles

A normal reversal in RIVR of TA and M generally has been reported to occur between early mixed dentition and late mixed dentition, reflecting increasing predominance of M activity.[12] This reversal reported was delayed one stage in patients with crowded dentition and whose occlusal contact area is smaller than in normal subjects.[12] In our study, activity of TA was predominant in I-G. Specifically, RIVR of M in I-G was significantly smaller than in II-G, as a result, no reversal in the RIVR of TA and M occurred. The predominant muscle used for mastication did not change from the temporalis to the masseter muscle in early mixed dentition in DMD subjects. We believe that this abnormality resulted from functional imbalance between masseter and temporalis muscles, and was related to the marked decrease in occlusal contact due to poor congruity between dental arches. This interpretation is supported by reports of a decrease in the integral value and shortening of the silent period of the masseter muscles in DMD patients.[20,31] It is supported by a report that skeletal open-bite was related to decreased strength in vertically oriented muscles in the posterior mandibular area.[32]

In normal subjects, DR of discharge generally is considered to decrease with increasing dental stage.[12] Another study has shown shortening in both DR and IT in normal subjects.[33] The generally accepted explanation is that because chewing efficacy in mastication is low in early dental stages, DR is prolonged as compensation. As chewing efficacy increases with dental age, DR shortens. This view is supported by a report showing a longer DR to be associated with a firmer consistency.[34,35]

In DMD subjects, DR was prolonged at every dental stage beyond that seen in control data. This prolongation may result from decreased chewing efficacy from declining masticatory muscle function, decreased occlusal contact area. and decreased stability of occlusion due to open-bite. Indeed, prolongation of rhythm (DR, IT and D+I) in the masseter muscle as well as irregularity of rhythm has already observed in I-G in DMD subjects, and consistent with the findings of Hamada *et al*.[36] These findings indicated that in DMD patients, prolongation of rhythmic occurrence intervals and irregularity in the rhythm pattern further decreased the function of the masseter muscle and impair masticatory function.[20]

Our findings suggested that morphological imbalance between mandibular and maxillary dentition such as open-bite and cross-bite occlusion, which became manifest from II-S to III-S, is related to progression of systemic symptoms in DMD patients. They also suggest that malocclusions in DMD patients are related to disproportionate functional decreases in circumoral soft tissue and muscles. In particular, incongruities between functional changes in the tongue and muscle groups lateral to the dental arch especially the masticatory muscles, may be related to complex malocclusion. In addition, downward overgrowth of the lower jaw occurred due to functional decline in the masseter muscle in late mixed dentition. These disproportionate morphological changes in the dentition and the divergence of the jaws of DMD subjects begin at late mixed dentition or early permanent dentition. It could be demonstrated that these alternations in circumoral function and form were related with progression of systemic disease.

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