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Bite force and maxillofacial morphology in patients with Duchenne-type muscular dystrophy

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Abstract:

Purpose: The purpose of this study is to evaluate maxillofacial morphology and bite force in patients with severe Duchenne-type myodystrophy.

Subjects and Methods: The subjects were 24 men (average age, 21.5 years; range, 17-30 years) with Duchenne-type muscular dystrophy receiving treatment in National Ioh Hospital. Lateral and axial cephalograms were used to assess the morphology in this study. Furthermore, the maximum bite force on the first molar and the maximum mouth opening distance were measured.

Results:

The anterior open bite was visualized in most patients on the lateral cephalogram. The upper and lower arch lengths in the patients were significantly smaller than those in the controls ($P<0.05$). In contrast, the upper and lower arch widths in the patients were significantly larger than those in the controls ($P<0.05$). The maximum bite force and maximum mouth opening distance in the patients were significantly lower than those in the controls ($P<0.05$).

Conclusion:

These results appear to be very useful for improving the care and treatment of patients with Duchenne-type muscular dystrophy.

Key words: Duchenne muscular dystrophy, maxillofacial morphology, bite force

Duchenne muscular dystrophy (DMD) is an X-linked recessive disorder that affects 1 in 3500 live-born males. The affected gene has been located in the short arm of the X-chromosome at the Xp21.2 site¹, and its protein product has been identified and called dystrophin.² Dystrophin is absent or hardly detectable in cases of DMD.¹ The progress of the disease is, in general, extremely slow and the muscles involved become atrophic and weak. In most cases, the disease manifests itself in the first or second year of life as a clumsy, unsteady gait. The muscular proprioceptive reflexes disappear at an early stage, and 5 to 10 years after the onset of the disease, patients are unable to walk. Myotonic dystrophy has a primary distal distribution; the principal affliction is of the hands, forearms, lower legs, and the muscles of the jaws, neck, face, and eyelids. The course of the disease varies from individual to individual. The disease may involve the muscles very early in life, but the development of severe muscle weakness can appear much later, entailing a late diagnosis.³ When the disease becomes terminal, the muscular affection is generalized, breathing becomes insufficient, and most patients die even during the second decade of life, or at the latest at beginning of the third, from recurrent infections of the upper airways. The life expectancy of patients with DMD is low; most patients die of respiratory failure between the ages of 16 and 21 years.

On the other hand, the facial appearance of patients with myotonic dystrophy is characteristic, related to the weakness of muscles in the face and jaws. In patients with progressive muscular dystrophy, a transversal expansion of the dental arches in the upper and lower jaws is accompanied by a posterior crossbite, a reduction of overbite, an increased occurrence of medial diastema in the upper and lower jaws, and delayed dental

development. A diminishing bite force and an enlargement of the tongue, which is ascribed to pseudohypertrophy, were also found.⁴⁻⁷ Previous studies examined the maxillofacial morphology using lateral cephalograms and dental casts. Because lateral expansion of the dental arches is suggested, examination with axial (submento-vertex) cephalograms may be useful for assessing horizontal morphology, including the morphology of the condyles.

The purpose of this study was to evaluate the maxillofacial morphology and bite force of patients with severe Duchenne-type myodystrophy, using not only lateral cephalograms but also axial cephalograms.

Patients and methods

The subjects were 24 men (average age, 21.5 years; range, 17-30 years) with severe cases of Duchenne-type muscular dystrophy receiving treatment at National Ioh Hospital. According to the classification of functional severity staging system from the Ministry of Health, Labor, and Welfare, Japan (Table I), two cases were stage 4b, two cases were stage 5, two cases were stage 6, four cases were stage 7a, three cases were stage 7b, and eleven cases were stage 7c. Lateral and axial cephalograms were used to assess the morphology in this study. The following conditions were used for axial cephalograms (Fig. 1): the focus-to-film distance was 130 cm, the ear rod-to-film distance was set at 15 cm, the tube voltage was 80 kV, and the tube current was 50 mA. The images of the apex in the ear rods were connected with a line.⁸

The cephalograms were input into a computer with scanner (GT9500, Epson, Tokyo, Japan), and analyzed using a computer software (Cephalometric A to Z, Yasunaga Labo Com, Fukui, Japan). In the analysis of the lateral cephalograms, the Ricketts method was used and the data of subjects were compared with normal Japanese adult data of the computer soft.

The measurements in the lateral cephalogram were defined by Ricketts as follows.

Anterior (Ant) Cranial Base: the distance between the cross point (Basion-Nasion plane and Facial axis (Pterygoid-Gnathion)) and Nasion.

Post Facial Height (Ht): the distance between Gonion and the cross point (Frankfurt Horizontal (FH) plane and the perpendicular line to the FH plane across the most posterior point of pterygomaxillary fissure).

SN-Palatinal Plane: the angle between Sella-Nasion plane and Palatal plane (anterior nasal spine-posterior nasal spine)

Mandibular plane: the angle between FH plane and Mandibular plane.

Total Facial Height (Ht): the angle between Basion-Nasion plane and Mandibular plane.

Convexity: the distance between A point and Facial plane (Nasion-Pogonion).

Lower Facial Height (Ht): the angle between ANS, XI (the center point of Ramus) and PM (Protuberane menti; Supra pogonion)

Lower Lip E-line: the distance between lower lip and esthetic-line.

The measurements in the axial cephalogram were defined as follows (Fig. 2):

Condylar width: the distance between the most lateral points of bilateral condyles;

Condylar length: the distance between the most medial point and the most lateral point;

Arch length: the distance between the line connected the most distal points of bilateral first molars and the mid-point of medial edges of bilateral incisors;

Arch width: the distance between the most distal points of bilateral first molars.

The maximum mouth opening distance (MMO) was calculated as (the distance between upper and lower incisors + overbite).

Furthermore, maximum bite force at the right first molar was measured with MPM-3000 (Nihon Kouden Co, Tokyo, Japan), and an occlusal force transducer with 17 mm in diameter at the end and a block 1 mm high and 3 mm in a diameter located the centre. The subjects were asked to bite on the block as hard as possible, while maximum digital readouts were measured and displayed in kg. Measurements of maximum force are dependent on the effort of the subject, and can be influenced by motivation, pain, and anxiety, among other factors. Thus, it is very important to check the reproducibility of these methods. So, the average bite force was used from the data of 3 times trial records.

The data of subjects in cephalograms, MMO and occlusal force were compared with control subjects, who were 11 healthy volunteer men (21.3 ± 3.5 years old) with normal skeletal occlusion.

Statistical analysis

Data from the cephalograms, occlusal force measurements, and maximum mouth opening distances were statistically analyzed with StatView™ version 4.5 software

(ABACUS Concepts, Inc., Berkeley, CA, USA). Differences between groups were analyzed by non-paired comparison using the Mann-Whitney U test. Differences were considered significant at $P < 0.05$.

Results

SNA in patients was significantly larger than that in the controls ($P < 0.05$), however, there was no significant differences in SNB. Anterior cranial base in patients was significantly shorter than that in the control ($P < 0.05$). These showed that the Nasion point located more posterior position in the patients.

Interincisor overbite in 23 of the 24 patients was less than 0 mm. The post facial height, total facial height, lower facial height, and lower lip E-line in patients were higher than those in the controls on lateral cephalogram analysis ($P < 0.05$). From these results, all patients showed an anterior open bite (Table 2).

On the other hand, there were no significant differences between patients and control subjects in condylar length and condylar width on axial cephalogram analysis (Fig. 3, Table 2).

The upper and lower arch widths of the patients were significantly greater than those of the controls ($P < 0.05$). In contrast, the upper and lower arch lengths were significantly shorter than those of the controls ($P < 0.05$) (Fig. 4, Table 2).

The occlusal force of the patients was significantly lower than that of the controls ($P < 0.05$) (Fig. 5A, Table 2).

No significant difference between patients and controls was found regarding maximum mouth opening distance, although that of patients tended to be slightly less (Fig. 5B, Table 2).

Discussion

Clinical studies have shown a correlation between bite force and facial morphology.⁹⁻¹¹ Subjects with a strong bite force also had a more anteriorly inclined mandible, smaller anterior heights, greater posterior facial heights, and a smaller gonial angle than subjects with a weak bite force. These findings support the theory that the form of the face partly depends on the strength of the muscles. However, Throckmorton et al.¹² stated that the low bite force in persons with long faces might be a result of the geometric arrangement of the lever system of the jaw. Thus, according to this view, it is the muscular function that is influenced by the skeletal form.

In patients with DMD, the frequency of malocclusion was very high.^{10,13} Furthermore, the masticatory muscles in these patients had been affected by the disease, as indicated by the low biting force compared with that in healthy controls or that found in previous studies.^{14,15}

The involvement of these muscles may have caused a lowering of the mandible, due to either gravitation or the activity of possibly less involved suprahyoid muscles.¹⁶ Lowering of the mandible can, in turn, affect the tongue position and head posture. Thus, a new situation is established transversely around the teeth. The lowered tongue is not in a position to counterbalance the forces developed during the lowering of the mandible by the

stretched facial musculature. This may affect the position of the teeth transversely, decreasing the width of the palate and causing a crossbite.¹⁵ In our study, similar results were obtained. However, regarding maximum mouth opening distance, there was no significant difference between the patients and the controls. This suggested that reduction of muscular power could not completely inhibit the components that function in opening the mouth, including the suprahyoid muscles and temporomandibular joint.

Patients with DMD usually have recurrent mandibular dislocation, which is not painful and easily returns to a closed position.¹⁷⁻¹⁹ Zanoteli et al.²⁰ reported that mild bone abnormalities, including changes in the shape and contour of the bone surface and sclerosis of the bone marrow, were frequently observed in a magnetic resonance imaging study of patients with myotonic dystrophy. In this study, severe temporomandibular joint symptoms including dislocation were not found in all cases, although it was impossible to examine the TMJ structure of patients in detail with only cephalograms. On the other hand, axial cephalograms showed that dental arch width was expanded, but condylar width and condylar length were normal. This suggested that lateral expansion in the alveolar process was present, uninhibited by volume growth of the mandible, including the TMJ. The lowered position of the mandible, in combination with the decreased biting forces, may permit an overeruption of the teeth.¹¹ In this case, the palatal vault height is possibly increased because of the overeruption, and the mandible rotates posteriorly, causing an increased angle between the mandibular and palatal planes. The increased frequency of angle class II malocclusion among patients with myotonic dystrophy may be attributable to the posterior rotation of the mandible. In our study, the class II tendency was not found.

On the other hand, despite the reduction in overbite in patients who have progressive muscular dystrophy of the Duchenne type, Eckhardt and Harzer²¹ did not observe any such increase in lower facial height compared with upper facial height, as described in patients who have a myotonic dystrophy. This suggests differing specific affection patterns for myotonic dystrophy. However, the subjects in their study had a mean age of 11.7 years, younger than subjects in other reports including our study. Severity of disease and age of the patients should be evaluated for studies to become comparable.

The management of DMD is progressing so that patients can live longer. An increased understanding of the functional and morphological changes of the oral and maxillomandibular regions may improve the oral care of these patients to further enhance their quality of life.

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Figure Legends

Figure 1. Cephalograms of patients with Duchenne-type muscular dystrophy.

Figure 2. Trace of axial cephalogram and measurements.

Figure 3. Comparison between the group of patients with Duchenne-type myodystrophy and the control group.

A: Condylar width. B: Condylar length. *, statistically significant difference: $P < 0.05$

Figure 4. Comparison between the group of patients with Duchenne-type myodystrophy and the control group. *, statistically significant difference: $P < 0.05$

Figure 5. Comparison between the group of patients with Duchenne-type myodystrophy and the control group.

A: Bite force. B: Maximum mouth opening distance. *, statistically significant difference: $P < 0.05$

Table 1. Grade of functional severity of myotonic dystrophy.

Table 2. Results of cephalometric analysis, MMO and bite force. *, statistically significant difference: $P < 0.05$

Fig. 1 Cephalograms of Duchenne type muscular dystrophy patients

Fig. 2 Tracing of axial cephalogram and measurements

Fig. 3 Comparison between Duchenne type myodystrophy group and control
A: Condylar width, B: Condylar length. *, statistically significant difference: $P<0.05$

Fig. 4 Comparison between Duchenne type myodystrophy group and control *, statistically significant difference: $P < 0.05$

A

B

Fig. 5 Comparison between Duchenne type myodystrophy group and control

A: Bite force, B: Maximum mouth opening distance. *, statistically significant difference: $P < 0.05$

Grade		Standard of motion	
1	possible to walk	possible to step stairs	a) without hand support b) with hand support to one's knee
2	possible to walk	possible to step stairs	a) with a handrail b) with a hand rail and one hand support to one's knee c) with handrail of both sides
3	possible to walk	possible to step stairs	possible to stand up from seated position a) nothing to support b) with something to support
4	possible to walk		a) longer than 5 m without help b) with support 1) a walker 2) a handrail 3) someone's hand
	impossible to stand up from seated position		
5	impossible to walk	possible to crawl on all fours and move	
6	impossible to crawl on all fours, but possible to crawl on elbows		
7	impossible to move		a) possible to move by oneself with a wheelchair b) impossible to move by oneself with a wheelchair, possible to keep seated position c) impossible to move by oneself with a wheelchair, impossible to keep seated position
8	bedridden and need maximum care		

Table 1. The grade of functional severity of myotonic dystrophy

		DMD		Control		
		Average	SD	Average	SD	
SNA	(dg)	85.5	3.3	80.7	4.3	*
SNB	(dg)	79.6	2.9	77.9	4.0	
ANB	(dg)	5.9	3.6	2.8	1.9	*
Ant Cranial Base	(mm)	58.8	3.6	68.8	3.3	*
Post Facial Ht	(mm)	74.6	7.9	80.7	3.9	*
SN-Palatinal Plane	(dg)	7.7	6.0	8.7	4.8	
Mandibular Plane	(dg)	38.6	7.4	27.7	5.3	*
Total Facial Ht	(dg)	71.5	7.2	61.8	5.3	*
Convexity	(mm)	8.6	5.4	3.2	2.9	*
Lower Facial Ht	(dg)	60.3	7.4	51.3	3.8	*
Interincisor Angle	(dg)	121.6	16.5	121.6	6.6	
Incisor Overjet	(mm)	3.0	3.8	4.6	1.2	
Incisor Overbite	(mm)	-8.9	8.3	1.1	1.3	*
Lower Lip E-Plane	(mm)	11.1	7.6	1.1	2.0	*
Condylar width	(mm)	129.8	6.9	125.6	8.0	
Condylar length	(mm)	17.4	4.2	18.5	2.9	
Upper arch length	(mm)	43.6	4.3	47.8	5.1	*
Upper arch width	(mm)	54.2	4.3	46.6	3.3	*
Lower arch length	(mm)	36.4	6.6	41.5	3.1	*
Lower arch width	(mm)	56.2	7.5	46.2	3.2	*
MMO	(mm)	36.9	12.5	41.0	8.0	
Bite force	(Kg)	8.1	7.4	52.2	15.4	*

Table. 2 The results of cephalometric analysis, MMO and bite force