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Developmental changes in craniofacial morphology in subjects with Duchenne muscular dystrophy

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SUMMARY Lateral cephalometric radiographs of 35 Japanese male patients suffering from Duchenne muscular dystrophy (DMD) were taken longitudinally from 10 to 20 years of age. Eighteen landmarks were placed and 15 angles and four linear distances calculated. Profile diagrams (profilograms) were produced to analyse changes in craniofacial morphological growth in the DMD subjects. The measurements were then compared with Japanese standards.

In young patients with DMD, compared with the controls, the following were observed: a large gonial angle; clockwise rotation of the mandible; short sagittal length of the cranial base and protrusion of the upper incisors. In adult patients, the maxillary alveolus and the upper incisors were protruded, compared with the controls. Overbite in DMD subjects also showed a tendency to decrease. In the controls, mandibular growth direction tended to be straight down and forward, while in patients with DMD, the growth direction was down until approximately 16 years of age and, after that, a forward vector of growth was apparent. As a result, the tendency towards a clockwise rotation of the mandible in the adults was less than in the young patients.

These findings showed that DMD significantly affects craniofacial morphology.

Introduction

Duchenne muscular dystrophy (DMD) is a severe form of progressive muscular dystrophy. It is an X-linked recessive disorder that affects 1 in 3500 live born males (Weller *et al.*, 1997). The affected gene has been localized in the short arm of the X-chromosome at the Xp21.2 site and its protein product has been identified as dystrophin. Dystrophin is absent or hardly detectable in DMD (Hoffman *et al.*, 1987; Weller *et al.*, 1997).

In most cases this disease is recognized at 1 or 2 years of age due to a clumsy, unsteady gait. The muscular proprioceptive reflexes disappear at an early stage, and 5 to 10 years after the onset of the disease the patients become wheelchair-bound. Initially, the gluteofemora, the quadriceps, and the adductors are affected, and later the arm muscles, the shoulder girdle, and eventually the facial muscles. Terminally, the muscular affection is generalized, breathing becomes insufficient, and most patients die during their teens or early twenties, from recurrent upper airway infections.

It is well known that specific malocclusions, for example an open bite, are found in patients with DMD (Figure 1). There is also general agreement that the widths of the upper and lower dental arches in DMD patients are greater than in controls, the differences being more pronounced in the mandible than in the maxilla (Futterman, 1940; Cohen, 1975; Nakashima *et al.*, 1984; Nakata *et al.*, 1984; Stenvik and Stohhaug, 1986) and this can lead to development of a posterior crossbite (Ghafari *et al.*, 1988; Erturk and Dogan, 1991). The length of the dental arches in DMD subjects is smaller than in controls. In addition, transverse overdevelopment and sagittal shortening of the dental arch leads to a reduction in overbite and overjet (Eckardt and Harzer, 1996). An anterior open bite (AOB) has also been reported in patients with DMD, but this is less frequent than a posterior crossbite (Manhartsberger *et al.*, 1987; Ghafari *et al.*, 1988). Studies by White and Sacker (1954), Ghafari *et al*. (1988), Watanabe *et al*. (1990) and Erturk and Dogan (1991) suggested that one of the causes of the malocclusions may be enlargement of the tongue, which has been ascribed to pseudohypertrophy. On the other hand, an electromyography study by Hara *et al*. (2002) suggested that the time lapse between atrophy of those muscles responsible for mouth-opening and mouth-closing might be causative factors in the development of malocclusions. Therefore, it appears that there is no agreement on what causes the specific dental and craniofacial changes in DMD and there are few reports on the longitudinal changes.

The purpose of this study was to analyse the influence of DMD on the craniofacial morphology and occlusion, paying particular attention to the developmental changes, by using longitudinal lateral cephalometric radiographs.

Subjects and methods

The subjects were 35 male patients who had lateral cephalometric radiographs taken longitudinally from 10 to 20 years of age. All patients were under treatment at

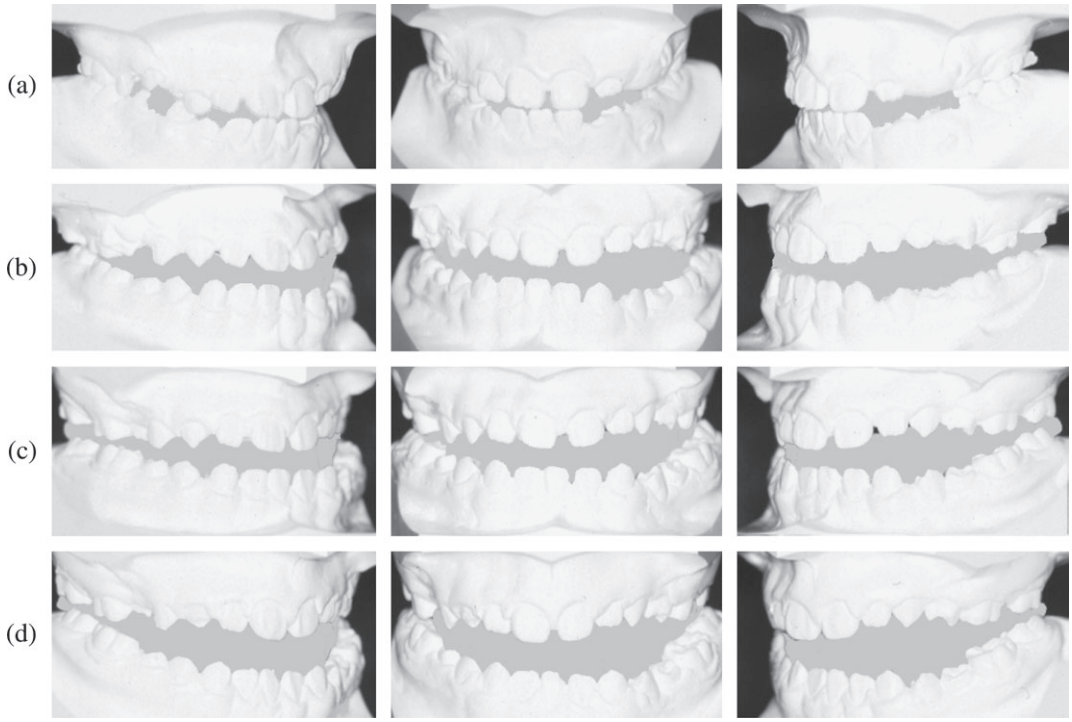

Figure 1 Models of a subject with Duchenne muscular dystrophy at (a) 8, (b) 11, (c) 14, and (d) 17 years of age.

Nishibeppu National Hospital, and ethical approval and co-operation for the research was given by that hospital. The study was mixed longitudinal, which is a research technique utilized to determine the changes associated with ageing or development. Information is obtained continually and longitudinally from a group of subjects, and the data are then sorted by age. Each group of data is averaged and rearranged in order of age and the averages are regarded as longitudinal.

As a result of transfers to other hospitals, deaths, and the difficulties encountered in taking usable cephalograms, the number of radiographs was reduced and was not evenly distributed between the different age groups. Therefore, cephalograms were not available longitudinally for each subject and this resulted in the use of a mixed-longitudinal analysis. In addition, the number of radiographs at 11 and 17 years of age was so small that a decision was made to exclude these time points.

The age distribution and number of lateral cephalometric radiographs is shown in Figure 2. The mean observation period was 4.7 years. None of the subjects had been prescribed immunosuppressants or steroid-based drugs which could have had an effect on the developing skeletal structures. Eighteen landmarks were placed on the cephalograms, and 15 angles and four linear measurements recorded (Figure 3a,b). Japanese male standards, at 13 and 20 years of age (Japanese Society of Pediatric Dentistry, 1995; Nagaoka and Kuwahara, 1993) were used as controls.

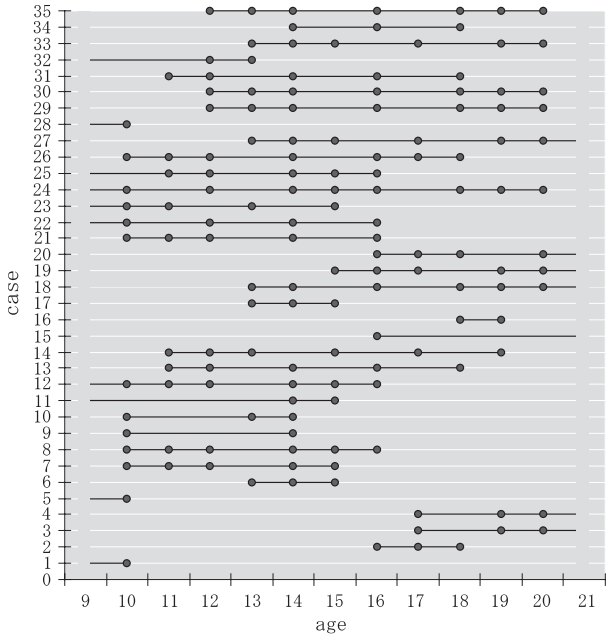

Figure 2 Age distribution for the cephalograms. The longitudinal axis represents the numbers of cases and the transverse axis ages.

The measurements were evaluated statistically and the DMD data compared with the controls.

In order to represent the growth changes visually, sella was used as the origin and a horizontal line passing through

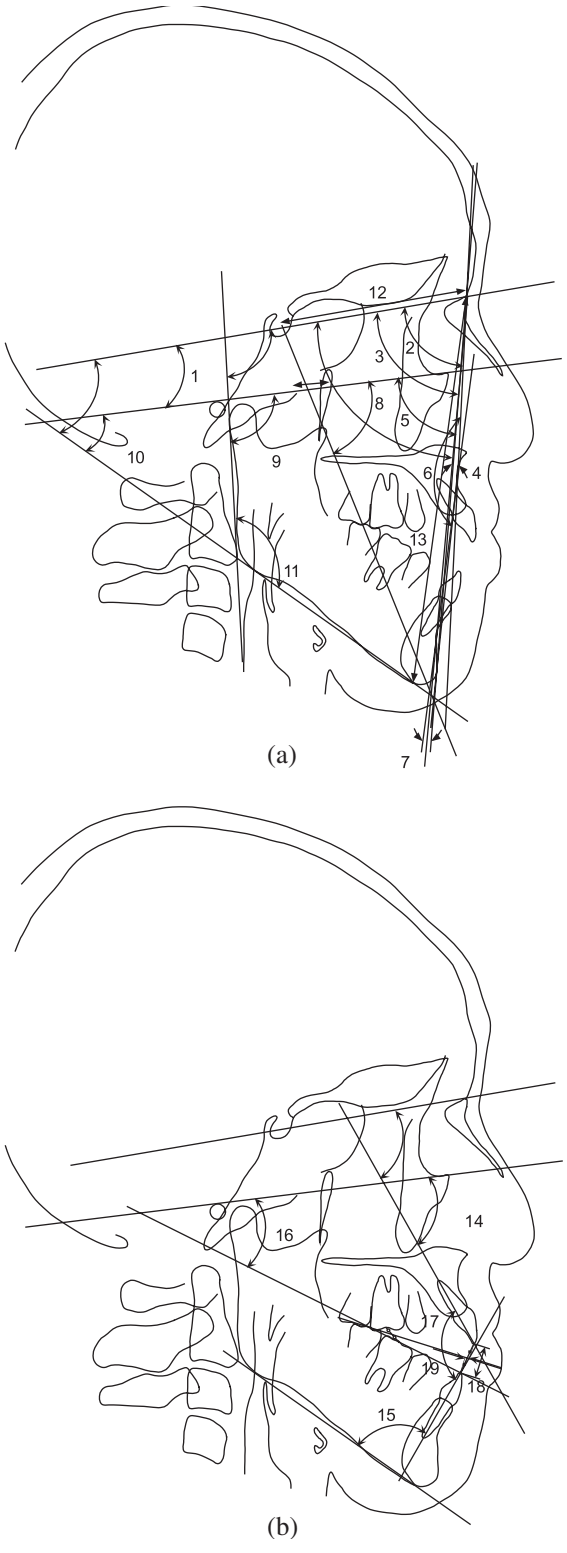

Figure 3 (a) Maxillo-facial angles and linear measurements: 1. FH–SN, 2. SNA, 3. SNB, 4. ANB, 5. FH–Facial, 6. Convexity, 7. AB-Facial, 8. Y–axis, 9. FH-Ramus, 10. FH–Mandibular, 11. Gonial, 12. S–N, 13. N–Me. (b) Dento-alveolar angles and linear measurements: 14. FH–U1, 15. L1–Mandibular, 16. FH–Occlusal, 17. Interincisal, 18. Overbite, 19. Overjet.

sella and parallel with the Frankfort horizontal (FH) plane as the *X*-co-ordinate axis. A line perpendicular to the *X*-co-ordinate axis and passing through sella was used as the *Y*-co-ordinate axis. The landmarks S, N, Or, ANS, PNS, A, U1, L1, L6, B, Pog, Me, Go, and Ar were placed in order and the mean co-ordinates of each point were calculated. Profile diagrams (profilograms) were then produced (Figures 4a,b) and the profilograms for subjects from 10 to 18 years of age were superimposed on the Japanese standard profilograms obtained from 9 to 18 years of age (Sato *et al.*, 2003; Figures 5a,b).

Using the mean co-ordinates for each landmark at each age point, linear and quadratic equations were calculated to illustrate changes in growth. In order to determine which equation (linear or quadratic) was a better fit, the F values were compared. When the quadratic equations are defined as $Y = aX^2 + bX + c$, the inflection points of the co-ordinate are $(-b/ab, -b^2 + 4ac/4a)$. If the quadratic equations were a significantly better fit than the linear equations, these inflection points could be regarded as 'turning points' of growth direction. Therefore, for each age group of DMD patients, the distances between the mean co-ordinates of L6, B, Pog and Me and the associated inflection points on the quadratic regression growth curves were calculated. This suggested that the ages closer to the inflection points were 'turning points' in growth direction (Figure 6).

The differences between the measurements for the DMD group and the standards were analysed using the Student's *t*-test. In addition, the difference in fit of each equation, linear or quadratic, was evaluated by analysis of variance. Significance was set at $P < 0.05$.

Results

Analysis of developmental changes by angles and distances

The measurements for the DMD patients are shown in Table 1a,b. Statistical comparisons between the DMD group and the Japanese standards, at 13 and 20 years of age, were undertaken.

At 13 years of age, *Y*-axis and FH–occlusal ($P < 0.01$) and FH–mandibular and gonial angle ($P < 0.001$) were significantly greater for the DMD group than the Japanese standards, but other measurements were significantly smaller compared with the Japanese standards: FH–SN ($P < 0.05$), AB–facial, SN, and interincisal ($P < 0.001$). Subsequently, FH–SN, *Y*-axis and FH–occlusal decreased and statistical significance was lost by 20 years of age.

Using FH–SN as an example, the mean value in the DMD group was 5.2 degrees at 13 years of age but reduced to 4.9 degrees at 20 years, while in the control group the mean value changed from 7.7 degrees at 13 years to 6.2 degrees at 20 years of age. Therefore in the DMD group, the value was smaller than in the control group at 13 years, following which there was a reduction in value, but as that in the

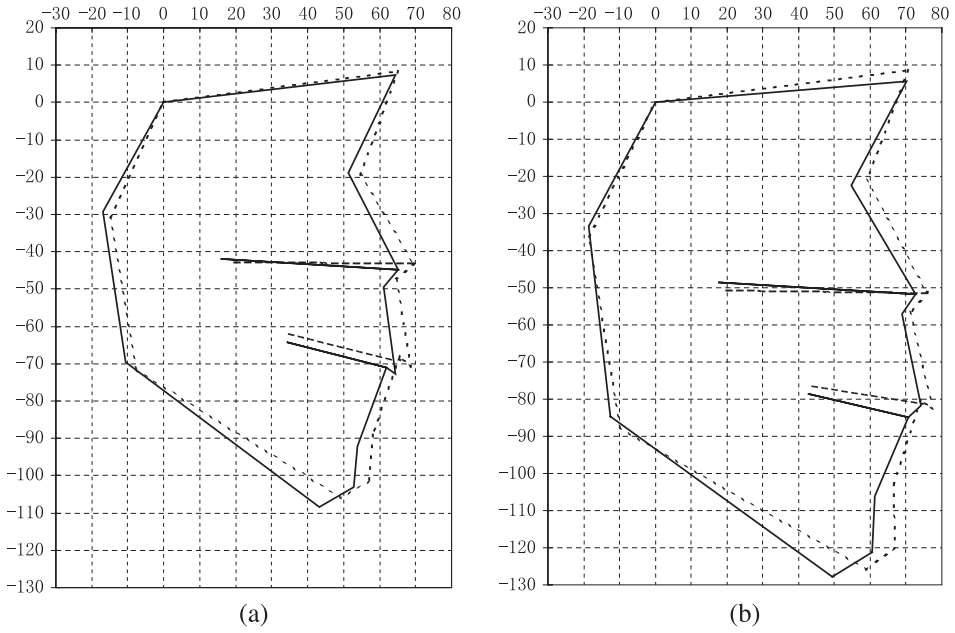

Figure 4 Superposition of profilograms of patients with Duchenne muscular dystrophy (black line) and same age standards (dotted line). (a) 10 years of age. (b) 18 years of age.

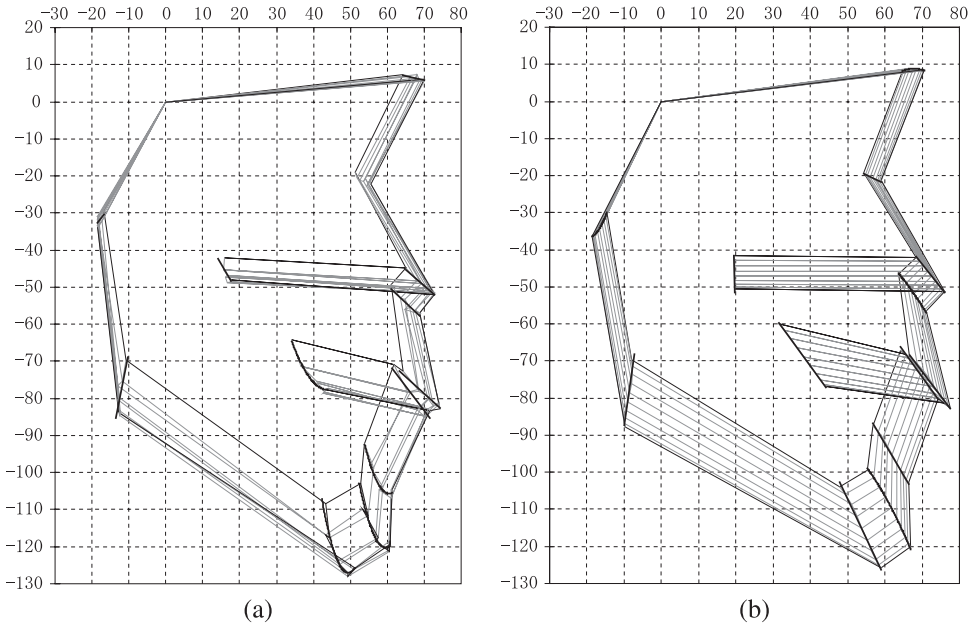

Figure 5 Superimposition of profilograms. (a) Patients with Duchenne muscular dystrophy from 10 to 20 years of age. (b) Controls from 9 to 18 years of age.

control group decreased by a greater amount, the statistical significance between the two groups was lost.

At 20 years of age, the measurements and statistical significance of FH–mandibular ($P < 0.05$) were also decreased. In contrast, FH–U1 in the DMD subjects tended to increase over time, with FH–U1 at 13 years of age not showing statistical significance, but at 20 years of age, the increase achieved significance ($P < 0.001$). At 13 years of age, differences in SNA and convexity between DMD subjects and the standards were not statistically significant. However at 20 years of age, SNA ($P < 0.05$) was significantly greater and convexity ($P < 0.05$) in DMD was significantly less than at 13 years of age (Table 1a).

In the 13-year-old DMD patients, the cranial base was smaller, the gonial angle was greater and the mandible was rotated clockwise to the FH plane, compared with the standards. However, at 20 years of age, the clockwise rotation of the mandible to the FH plane reduced, the

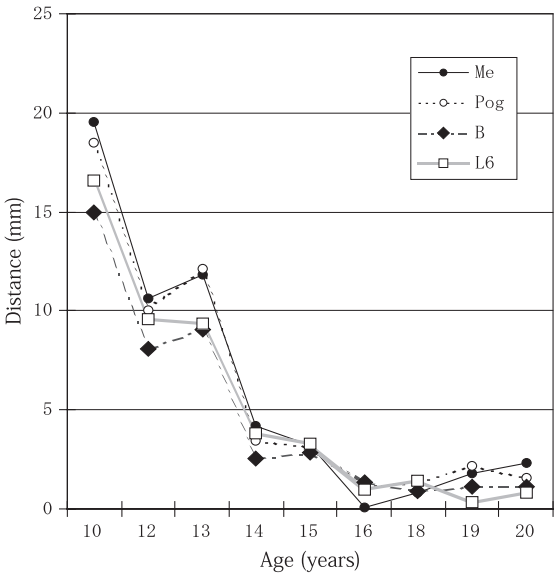

Figure 6 Distance between inflection point of quadratic regression curve and mean co-ordinates at each age.

maxillary alveolar bone developed forward and the maxillary anterior teeth were proclined, compared with the standards. In the DMD patients, the overbite was positive at 10 years of age but became negative by 12 years of age. Overbite reached a 'negative peak' between 15 and 18 years of age, and then increased again later (Table 1b).

Analysis of developmental changes by profilograms

The profilograms of the DMD patients and the control group standards were superimposed at 10 and 18 years of age (Figure 4). The gonial angle in the DMD group was already greater compared with the standards, the mandible was rotated downward and the profile was dolichofacial. This tendency was maintained at 18 years of age. The profilograms of DMD subjects from 10 to 20 years of age are shown in Figure 5a, and those of the standards from 9 to 18 years of age in Figure 5b.

The mean co-ordinates of each landmark were connected for age to show the changes during development. Based on that, linear and quadratic regression equations were

Table 1a Mean values and standard deviations of angles and distances in patients with Duchenne muscular dystrophy (DMD) from 10 to 20 years of age (except 11 and 17 years of age).

	DMD		DMD		DMD			Standard		DMD		DMD	
	Age 10, *n* = 13		Age 12, *n* = 15		Age 13, *n* = 14			Age 13, *n* = 25		Age 14, *n* = 22		Age 15, *n* = 12	
	Mean	SD	Mean	SD	Mean	SD		Mean	SD	Mean	SD	Mean	SD
Skeletal													
Angle (°)													
1. FH–SN	6.5	3.4	5.9	3.7	5.2	2.8	−	7.7	3.1	5.4	3.1	5.9	2.5
2. SNA	80.1	3.0	81.3	3.9	83.3	2.1		81.6	2.9	82.8	3.2	81.0	2.7
3. SNB	77.5	3.5	78.7	4.8	78.8	1.4		78.4	3.1	80.0	4.7	78.8	4.7
4. ANB	2.6	2.4	2.6	2.9	4.5	1.4		3.3	2.1	2.8	3.0	2.2	3.4
5. FH–facial	84.1	3.0	84.5	5.2	83.8	3.1		86.4	3.2	85.4	5.3	84.9	6.9
6. Convexity	174.8	5.5	174.5	6.8	170.4	3.4		173.6	4.8	174.6	7.1	176.0	8.7
7. AB–facial	0.9	1.2	0.7	1.6	1.2	1.9	−−−	5.3	3.3	1.0	1.6	0.8	1.7
8. *Y*-axis	65.5	3.0	66.6	5.5	67.1	3.7	++	63.2	3.4	66.7	5.5	66.7	7.0
9. FH–ramus	84.1	4.1	83.8	6.0	86.5	5.1		84.6	3.9	85.9	6.3	85.4	6.2
10. FH–mandible	34.2	4.2	35.8	8.9	34.0	5.3	+++	27.6	4.9	35.2	8.7	35.3	8.8
11. Gonial	130.2	5.1	132.0	7.5	127.5	3.3	+++	122.6	5.4	129.3	7.3	129.9	5.8
Distance (mm)													
12. S–N	64.9	2.4	66.9	2.8	66.8	3.8	−−−	70.5	2.8	68.0	3.7	69.4	4.6
13. N–Me	117.8	2.6	127.0	7.2	126.4	6.5		128.2	6.9	132.5	8.1	133.8	9.9
Dental													
Angle (°)													
14. FH–U1	116.7	4.7	118.9	5.9	116.3	7.2		115.4	7.2	118.7	8.1	122.4	9.1
15. L1–mandible	88.9	7.2	90.4	7.4	96.7	6.0		95.6	5.2	91.6	8.2	91.2	8.7
16. FH–occlusal	15.0	3.3	13.2	4.8	13.2	4.3	++	9.7	4.0	12.5	5.8	12.2	7.2
17. Interincisal	120.1	8.9	114.9	10.7	113.1	10.3	−−	121.4	7.4	114.6	9.6	111.1	13.8
Distance (mm)													
18. Overbite	1.7	2.5	−0.8	6.0	−0.4	3.4				−2.2	5.7	−3.6	5.0
19. Overjet	2.6	1.6	2.7	2.5	2.3	2.1				1.9	2.9	3.3	3.6

Mean values of cephalometric parameters in patients with DMD were statistically compared with those of standards at 13 and 20 years of age with an unpaired *t*-test.
+++ $P < 0.001$; ++ $P < 0.01$; + $P < 0.05$.
+++ denotes a greater value, −−− denotes a smaller value.

Table 1b Mean values and standard deviations of angles and distances in patients with Duchenne muscular dystrophy from 10 to 20 years of age (except 11 and 17 years of age).

	DMD		DMD		DMD		DMD			Standard	
	Age 16, *n* = 18		Age 18, *n* = 12		Age 19, *n* = 13		Age 20, *n* = 12			Age 20, *n* = 100	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD		Mean	SD
Skeletal											
Angle (°)											
1. FH–SN	4.7	3.7	4.6	3.7	4.6	2.4	4.9	2.6		6.2	2.8
2. SNA	83.7	4.2	84.6	4.2	84.0	2.9	83.9	3.0	+	81.4	3.6
3. SNB	81.4	5.4	81.6	5.9	81.3	4.2	80.6	3.4		79.6	3.9
4. ANB	2.3	3.2	3.0	3.5	2.7	2.7	3.3	2.9		1.8	1.6
5. FH–facial	86.3	6.4	86.4	8.3	86.2	5.8	85.9	5.5		87.4	2.8
6. Convexity	175.9	7.4	174.7	8.3	175.3	6.7	174.2	7.6	−	179.0	4.7
7. AB–facial	1.0	1.9	1.4	1.7	1.6	1.7	2.0	1.7	−−−	−3.0	2.8
8. Y-axis	66.0	7.2	66.0	10.1	65.6	6.1	65.4	5.5		63.3	2.8
9. FH–ramus	86.4	6.0	86.3	8.1	86.9	5.6	86.4	4.4		84.7	4.3
10. FH–mandible	33.9	12.3	33.5	17.2	32.2	8.2	31.4	6.9	+	25.6	5.6
11. Gonial	127.5	10.2	127.2	11.7	125.3	5.2	125.1	3.0	+++	112.4	6.0
Distance (mm)											
12. S–N	69.2	3.4	70.0	1.9	69.5	2.8	70.4	3.2	−−−	72.9	2.9
13. N–Me	135.3	11.9	136.1	14.5	133.5	6.8	133.6	8.4		136.0	5.2
Dental											
Angle (°)											
14. FH–U1	117.3	7.3	117.9	10.5	120.3	8.2	116.6	6.3	+++	111.2	5.2
15. L1–mandible	88.2	11.7	90.0	14.4	93.0	7.8	94.5	7.2		94.7	6.9
16. FH–occlusal	12.2	5.4	11.3	9.3	9.9	4.6	11.1	3.6		9.4	3.7
17. Interincisal	120.5	9.5	118.6	11.1	114.5	10.8	117.4	9.4	−−−	128.3	8.8
Distance (mm)											
18. Overbite	−2.9	7.5	−3.1	9.4	−1.7	5.0	−0.4	4.3			
19. Overjet	2.6	3.6	3.5	5.0	2.8	3.4	2.6	2.7			

Mean values of cephalometric parameters in patients with DMD were statistically compared with those of standards at 13 and 20 years of age with an unpaired *t*-test.
+++ $P < 0.001$; ++ $P < 0.01$; + $P < 0.05$.
+++ denotes a greater value, −−− denotes a smaller value.

calculated and the equations and the degree of fit of each (R^2 coefficient of determination) are shown in Table 2. For the standards, the R^2 values of the linear equations, except for N and PNS, exceeded 0.90. For landmarks L6, B, Pog and Me, the R^2 values were even higher, exceeding 0.99. The graphs tended to follow a straight line, and spread downward and forward as the ages increased. In DMD subjects at the same points (L6, B, Pog and Me) the quadratic equations were a better fit than the linear equations ($P < 0.05$), and therefore the graphs showed curved lines. As the coefficients on the quadratic terms of these quadratic regression equations were positive, these equations followed a convex downward curve. The distances between the mean co-ordinates of each landmark (L6, B, Pog and Me) at every age, and the inflection points on the regression curves, were calculated individually. The graph, in which the distances were established as the *Y*-co-ordinate axis and the ages from 10 to 20 years as the *X*-co-ordinate axis, is shown in Figure 6. The mean coordinates of each landmark approached the inflection points at approximately 16 years of age. The direction of development changed at 16 years of age, indicating that the anterior and lower part of the mandible grew downward initially and then forwards as the patients became older. In the control group, for the landmarks N, A, Pog and Ar, the quadratic equations were a better fit than the linear. The inflection points of the regression curves of the control group were not close to the mean co-ordinates of the measurement points and profilograms, thus it appears that these points were not related to age.

Discussion

With regard to the relationship between the perioral muscles and craniofacial morphology, Scotland and Rowland (1964), Gilroy and Meyer (1979), Swiman and Wright (1982), and Ghafari *et al.* (1988) reported that the mandibular position is mainly correlated with muscle elasticity, gravity, and intra-oral pressure, whereas craniofacial morphology is controlled by the contraction of the perioral muscles through sensory feedback. Moss and Rankow (1968), Subtelny (1970), and Kreiborg *et al*. (1978) suggested that weakened masseter and temporal muscles result in a lower resting

Table 2 Statistically significant differences between linear and quadratic regression equations in subjects with DMD and the control group.

	DMD								Control							
	Linear equation $y = ax + b$			Quadratic equation $y = ax^2 + bx + c$					Linear equation $y = ax + b$			Quadratic equation $y = ax^2 + bx + c$				
	a	*b*	R^2	*a*	*b*	*c*	R^2		*a*	*b*	R^2	*a*	*b*	*c*	R^2	
N	−0.25	23.41	0.473	0.00	−0.51	32.13	0.473		0.01	7.65	0.713	−0.07	8.88	−292.03	0.771	**
Or	−0.59	10.45	0.690	0.19	−21.00	555.35	0.779		−0.46	5.33	0.959	0.07	−8.18	223.97	0.990	
PNS	−1.70	−18.49	0.436	1.09	−37.91	282.13	0.488		−33.27	605.67	0.587	27.90	−1127.10	113.26	0.593	
ANS	−0.89	12.83	0.974	0.04	−6.91	220.68	0.988		−1.26	44.06	0.999	0.01	0.30	−12.30	0.999	
A	−0.92	5.33	0.844	0.10	−14.21	437.94	0.911		−1.44	45.45	0.996	−0.03	2.38	−83.24	0.998	**
U1	−0.91	−14.94	0.930	0.06	−8.87	261.99	0.966		−1.33	20.59	0.991	−0.03	3.00	−135.74	0.995	
L1	−1.31	8.37	0.873	0.13	−18.05	567.37	0.944		−1.41	24.55	0.992	−0.03	2.90	−124.81	0.996	
L6	−1.28	−22.80	0.916	0.14	−12.09	187.59	0.972	*	−1.37	−16.44	0.999	−0.00	−1.14	−20.74	1.000	
B	−1.55	−11.70	0.839	0.30	−36.51	998.76	0.951	*	−1.69	9.28	0.991	−0.02	1.32	−83.31	0.993	
Pog	−1.70	−18.04	0.811	0.33	−39.90	1069.00	0.930	*	−1.89	6.18	0.992	−0.04	3.18	−148.57	0.997	*
Me	−1.79	−37.33	0.744	0.40	−39.31	843.24	0.900	*	−2.09	−2.80	0.993	−0.03	1.63	−101.98	0.996	
Go	5.00	−17.86	0.511	3.84	96.05	519.73	0.663		6.86	−18.97	0.939	0.55	16.52	23.17	0.941	
Ar	1.23	−9.58	0.270	2.26	81.82	706.14	0.403		1.60	−7.07	0.966	0.28	11.03	70.92	0.996	**

*$P < 0.05$; **$P < 0.01$.

position of the mandible relative to the maxilla, and changes in the balance among the various muscles, due to atrophy and necrosis, influence the growth and development of craniofacial morphology and occlusion. Weijs and Hillen (1984) and van Spronsen *et al*. (1991) researched the relationship between the masticatory muscles and profile by computed tomography and reported that the activity of the masseter and internal pterygoid muscles affected facial morphology. Penarrocha *et al*. (1990) found that the functional conditions of soft tissues were closely related to the maintenance and development of the skeletal form (Ingervall and Thilander, 1974; Weijs and Hillen, 1984; Fukazawa and Mitani, 1985; Vilmann *et al*., 1985; van Spronsen *et al*., 1991, 1992). Therefore, it is suggested that atrophy of the perioral musculature in DMD is related to the age of onset and the severity of DMD, and can affect the skeletal form.

As regards changes in cranial morphology, Eckardt and Harzer (1996) and Cudny (1978) reported that the cranial base in DMD patients was short. In the present study, the S–N distance was also significantly smaller compared with the controls, thus reinforcing earlier findings. This suggests that muscular atrophy has an influence not only on the perioral region, but also on the cranial base.

Eckardt and Harzer (1996) reported that the activity of the muscles involved in closing the mouth, particularly the masseter muscle, was weakened by the age of 10 years, and that two years later, the activity of the orbicularis oris muscle was also weakened. Hara *et al*. (2002) suggested that this was due to the mouth-closing muscles performing isometric contraction against gravity, thus they bear heavier loads than those muscles leading to mouth opening, and are therefore more likely to atrophy. Nakashima *et al*. (1984) reported that the mandibular plane angle and *Y*-axis are increased in DMD patients, the mandible rotates in a postero-inferior direction, and the mandibular incisors become lingually inclined. Similar findings were noted in the 10-year-old patients in the current study, suggesting that the mouth closing muscles were beginning to atrophy, thus losing the ability to guide mandibular growth in the superior direction. Therefore, it seems likely that the postero-inferior rotation of the mandible and the dolichofacial morphology with an increased mandibular angle, noted at the age of 10 years, were caused by atrophy of those muscles involved in closing the mouth.

It was found that the direction of growth of the mandible changed from an inferior to an anterior direction around the age of 16 years. This may suggest that since the mouth-opening muscles weakened several years later than the mouth closing and orbicularis oris muscles, anterior growth of the mandible then dominated over downward growth. Furthermore, atrophy of all the striated muscles begins during this period, patients with DMD become wheelchair-bound, and changes in their head posture occurs. Therefore, changes in head posture may also be related to changes in mandibular growth direction.

Although an AOB is one of the malocclusion characteristics in DMD patients, positive overbites were still noted in patients aged 10 years. Thereafter, the overbites reduced and AOBs were noted in patients aged 15 and 16 years. Björk (1969) suggested that an AOB develops as the maxillary bone grows vertically, whereas the mandibular bone grows antero-posteriorly rather than vertically; therefore, the mandibular angle increases, setting the most posterior teeth as the centre of rotation. In their investigation

into the relationship between morphometric measurements and electromyographic findings, Hara *et al*. (2002) proposed that one cause of an AOB in DMD patients was mandibular postero-inferior rotation, which is caused by a combination of insufficient vertical growth of the mandibular ramus due to a decrease in the functionality of those muscles responsible for closing the mouth, as well as the extrusion of the maxillary molars. They considered that decreases in the activity of the mouth-closing muscles, such as the masseter, caused distortion of the vertical growth of the maxillo-mandibular bones and the posterior area of the dentition, thus resulting in an AOB.

On the other hand, as Ghafari *et al*. (1988) and Erturk and Dogan (1991) indicated, both pseudohypertrophy of the tongue and perioral muscle atrophy occur in DMD patients. As reported by Tamari *et al*. (1991), since pseudohypertrophy of the tongue may increase the width of the posterior part of the mandibular arch, this may result in a posterior crossbite and reduced incisor overlap. Perioral muscle atrophy also results in proclination of the maxillary labial segment, and consequently there is a reduction in overbite and an increase in overjet.

Kiliaridis and Katsaros (1998) reported that, although increased tongue pressure by pseudohypertrophy resulted in buccal expansion of the mandibular dentition toward the weakened masticatory muscles, the anterior mandibular teeth were not proclined as a result of being controlled by the orbicularis oris muscle which atrophies at a later stage. The findings in the present investigation, that the mandibular anterior teeth were not proclined, supports this theory.

The results of the present study also suggest that when the activity of the muscles responsible for mouth closure are weakened, tension in the other perioral muscles becomes relatively stronger, thus inducing downward traction on the mandible, and in particular causing downward growth of the anterior region of the mandible. Thereafter, when the strength of the mouth-opening muscles declines, and the downward mandibular traction ceases, the mandible begins to grow in an anterior direction. Furthermore, pseudohypertrophy of the tongue, poor head posture (such as a backward tilt of the head and neck), and oral habits such as mouth breathing are factors which are likely to be involved in morphological changes in the head and neck region (Ingervall and Thuer, 1988; Iino *et al.*, 2001; Solow and Kreiborg 1977). For example, when the muscles in the neck region are weakened and become unable to support the head, the patient begins to have a compensatory posture with the head leaning backwards. The present findings suggest that the balance of soft tissue, such as the muscles, the tongue, and posture are not only necessary for maintaining the skeletal form, but also greatly influence growth and development. This is an important finding in the field of orthodontics. Further research is, however, necessary to clarify the time of onset of atrophy of the mouth closing, orbicularis oris, and mouth opening muscles, and the deterioration of muscular strength. Furthermore, the time lapse between the decline in muscle functionality and morphological changes warrants further study.

Conclusions

The cranial base was shorter in subjects with DMD than in the control group and most of the DMD patients were dolichofacial. From childhood, the angle of the mandible was greater, the mandible rotated downwards and backwards, the overbite was reduced and the upper anterior teeth were proclined, compared with normal standards. However, downward and backward rotation of the mandible reached a peak at approximately 16 years of age, after which the growth pattern tended to improve. The results of this study show that craniofacial growth and occlusion are influenced by DMD.

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