

# Early Orthodontic Treatment and Long-term Observation in a Patient with Morquio Syndrome

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**Abstract:** Early orthodontic treatment and long-term observation in a patient with Morquio syndrome were demonstrated. To date, there are no case reports describing orthodontic treatment in such a patient. The patient showed spaced maxillary and mandibular arches with a tongue thrust habit at age seven years eight months. She also showed a protruded upper lip, labial inclination of the upper and lower anterior teeth, and thin enamel. The tongue thrust habit disappeared after the application of a removable orthodontic appliance. The spaced dentition in the upper and lower arches improved after treatment with a fixed appliance with closing loop arch wire ( $0.017 \times 0.025''$ ) and bands on the upper and lower molars. We did not use an edgewise appliance because of the thin enamel. The protruded upper lip and labial inclination of the upper and lower anterior teeth were also improved after treatment, but optimal intercuspation of the teeth was not achieved. However, optimal intercuspation of the teeth was achieved after long-term observation and the masticatory function was improved. It was suggested that early orthodontic treatment could improve the malocclusion in a patient with Morquio syndrome and that improvement of masticatory function could be achieved during a long-term retention period. (*Angle Orthod* 2005;75:881–887.)

**Key Words:** Early orthodontic treatment; Long-term observation; Morquio syndrome

## INTRODUCTION

Morquio syndrome (mucopolysaccharidosis IV) was first reported by Morquio<sup>1</sup> and Brailsford<sup>2</sup> and is a genetic disorder characterized by the lack of *N*-acetylgalactosamine-6-sulfatase activity.<sup>3</sup> This syndrome

shows an autosomal recessive heredity, and it affects one in 40,000 live births.<sup>4</sup> This syndrome is also characterized by short stature, short neck, pigeon breast, corneal opacities, spinal deformity, knock knees, deafness, muscle weakness,<sup>4</sup> and excessive urinary excretion of keratosulfate.<sup>5</sup> In the dento-orofacial area, the syndrome is characterized by a flat nasal bridge, flared alae nasae, broad mouth,<sup>4</sup> labially inclined upper incisor, spaced arch,<sup>6–8</sup> thin enamel, and cracks and pits of the enamel.<sup>6–12</sup> Therefore, it can be speculated that patients with this syndrome often need orthodontic treatment. However, there are no case reports in which orthodontic treatment was performed in such patients.

Mastication is an important function affecting the quality of life (QOL) in humans. Previous studies have shown that masticatory function is lower in patients with malocclusion compared with that in subjects with good occlusion.<sup>13–15</sup> Therefore, it is suggested that the correction of malocclusion by orthodontic treatment may be useful for the improvement of masticatory function, particularly in patients with Morquio syndrome. This study demonstrates early orthodontic treatment and long-term observation in a growing patient with Morquio syndrome.

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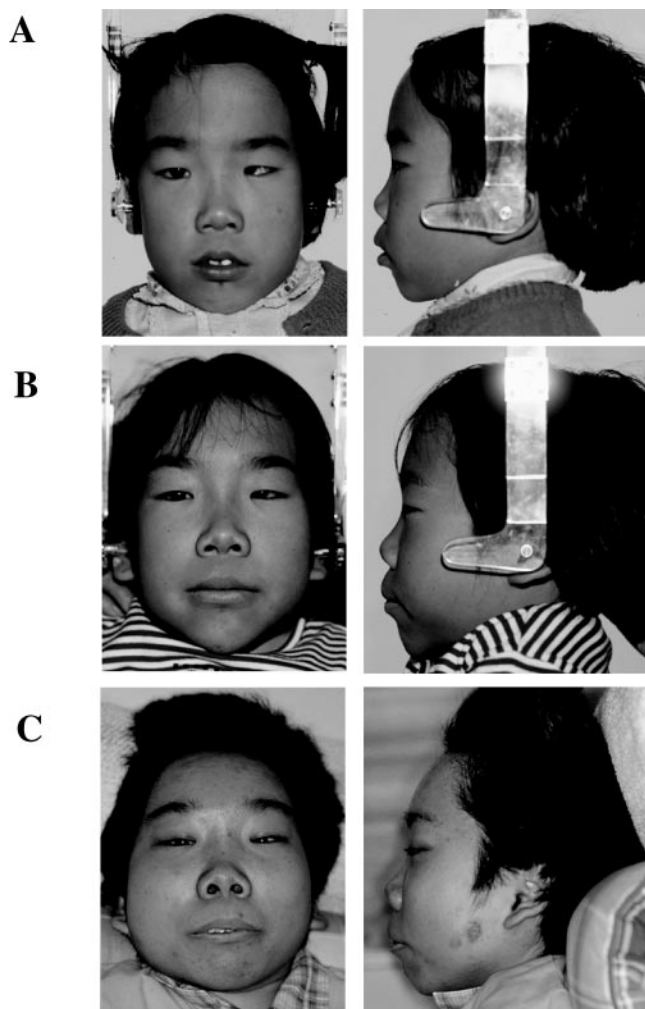
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**FIGURE 1.** Facial photographs. (A) Initial stage (seven years eight months). (B) Postactive treatment stage (12 years nine months). (C) Postretention stage (26 years five months). Left: frontal view; right: lateral view.

### Case summary

The patient was a seven-year eight-month-old Japanese girl who was diagnosed as having Morquio syndrome. Her chief complaint was spacing in the upper and lower dentition. She showed a short stature, short neck, pigeon breast, corneal opacities, and knock knees. She also showed a convex and symmetrical profile. She demonstrated protrusion of both the upper lip and the maxillary incisors (Figure 1A).

The lateral cephalometric radiograph showed a long cranial base length (S-N 74.8 mm, Z-score +4.5). The anteroposterior relationship between maxilla and mandible was skeletal Class I (ANB angle 4.0°, Z-score +0.2). However, both the maxilla and mandible were located in more posterior positions (SNA angle 72.6°, Z-score -2.9; SNB angle 68.6°, Z-score -3.6) than normal. The mandibular plane angle was high (Mp-SN angle 46.2°, Z-score +2.0). Upper and lower incisor

**Table 1.** Cephalometric Analysis

Measurements	Initial Stage (7 y 8 mo)		Postactive Treatment Stage (12 y 9 mo)	
	Value	Z-Score <sup>a</sup>	Value	Z-Score <sup>a</sup>
Angular (°)				
ANB	4.0	0.2	5.4	1.1
SNA	72.6	-2.9	74.4	-1.9
SNB	68.6	-3.6	68.9	-2.1
Mp-SN	46.2	2.0	47.8	2.3
U1-FH	115.7	1.0	108.9	-0.4
L1-Mp	110.4	3.3	108.3	3.0
Linear (mm)				
OJ	3.5	0.7	5.0	1.9
OB	1.0	-0.8	3.0	-0.4
N-S	74.8	4.5	77.5	3.1
U1-NF	32.0	2.6	32.7	1.5
L1-Mp	43.1	1.9	44.7	1.1
U6-NF	19.0	1.0	22.7	0.3
L6-Mp	31.3	1.2	34.3	1.5

<sup>a</sup> Z-score was calculated as (value - norm)/1 SD, using norms and SDs of mean Japanese females according to the corresponding age.<sup>16</sup>

teeth showed a labial inclination (U1-FH angle 115.7°, Z-score +1.0; L1-Mand.pl angle 110.4°, Z-score +3.3) and extrusion (U1-NF 32.0 mm, Z-score +2.6; L1-Mand.pl 43.1 mm, Z-score +1.9). The upper and lower molar teeth also showed extrusion (U6-NF 19.0 mm, Z-score +1.0; L6-Mand.pl 31.3 mm, Z-score +1.2; Table 1, Figure 6). Furthermore, the patient had a tongue thrust habit.

She showed an Angle Class I malocclusion with spacing in the upper and lower dentition. Overjet and overbite were 3.9 and 0.9 mm, respectively. The maxillary dental midline corresponded to the mandibular midline. The buccal cusps of the molars were tapered (Figures 2A, 3A, and 4A).

The panoramic radiograph indicated a normal number of permanent tooth germs, and four third molar buds could be observed. The tooth enamel was considered thin (Figure 5A).

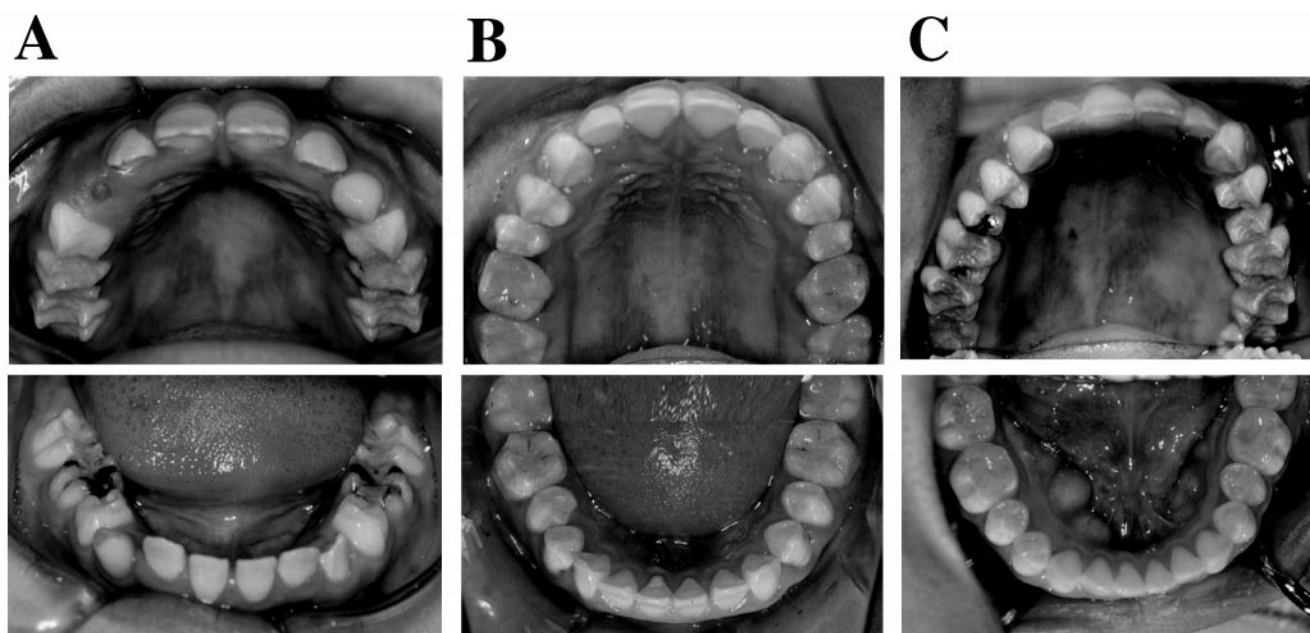
### Diagnosis and treatment plan

The patient was diagnosed as having an Angle Class I malocclusion with dentoalveolar bimaxillary protrusion, upper and lower spaced arches, skeletal Class I, and high mandibular plane angle.

The orthodontic treatment was planned as follows: (1) oral hygiene improvement by oral hygiene instruction, (2) prevention of tongue thrust habit by using removable appliance, (3) space closure in the upper and lower dentition using a removable or fixed orthodontic appliance, and (4) long-term retention.

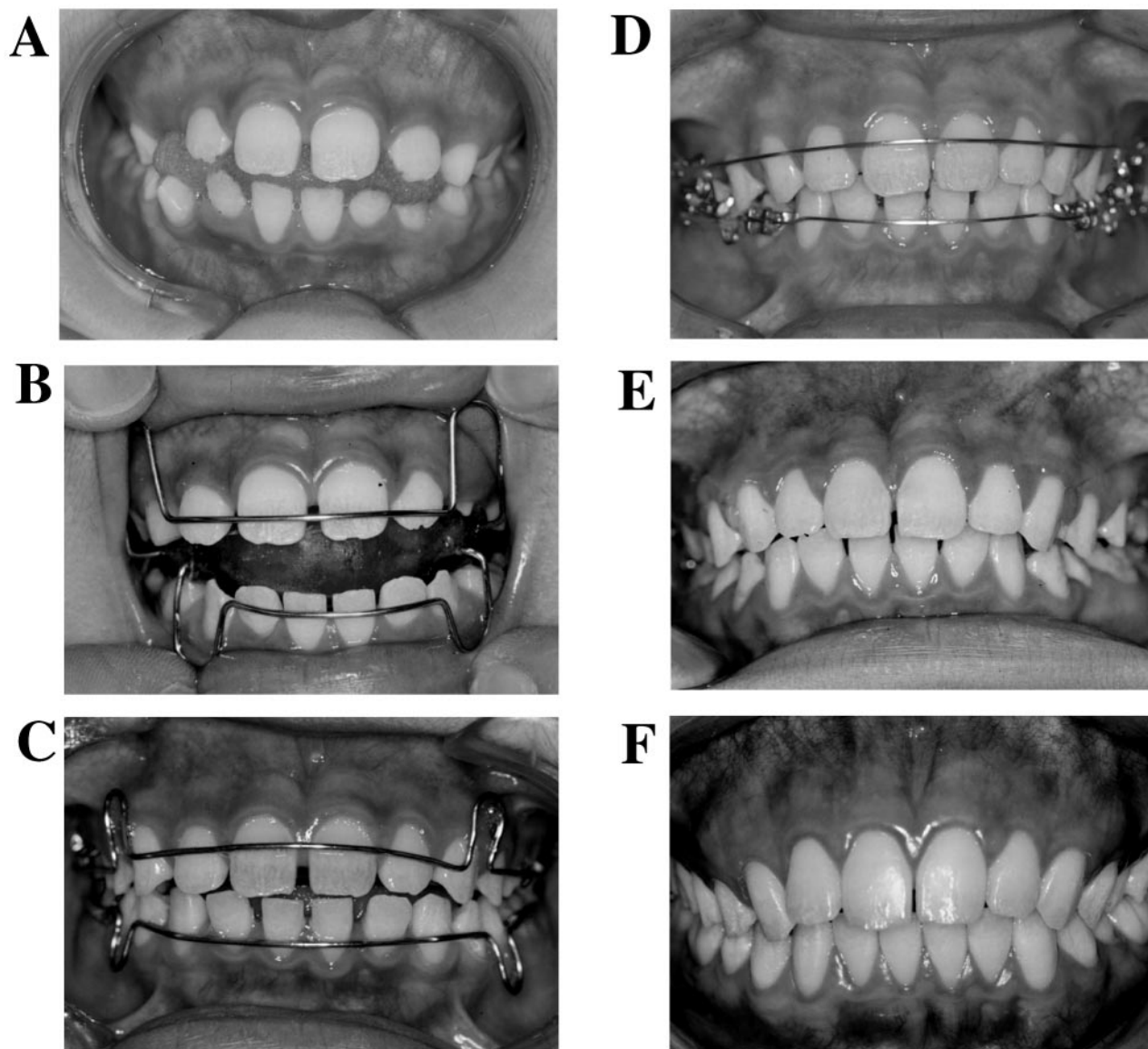


**FIGURE 2.** Intraoral photographs. (A) Initial stage (seven years eight months). (B) Postactive treatment stage (12 years nine months). (C) Postretention stage (26 years five months). Left: lateral view on the right side; center: frontal view; right: lateral view on the left side.



**FIGURE 3.** Intraoral photographs. (A) Initial stage (seven years eight months). (B) Postactive treatment stage (12 years nine months). (C) Postretention stage (26 years five months). Upper: occlusal view of upper dentition; lower: occlusal view of lower dentition.





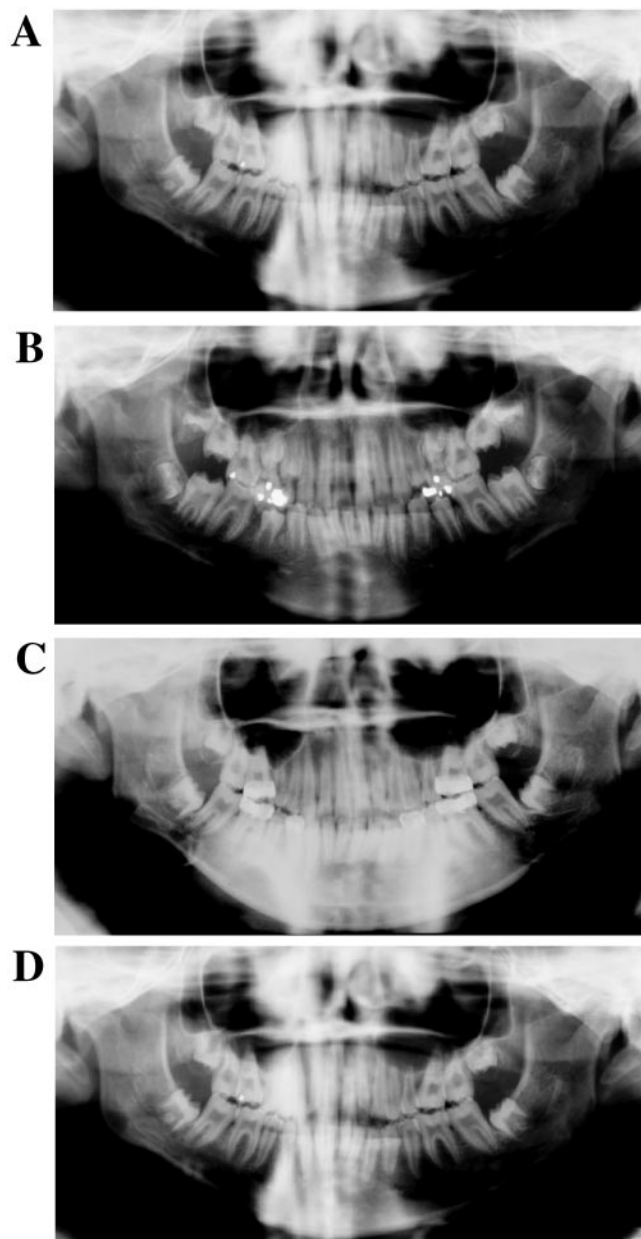
**FIGURE 4.** Intraoral photographs. (A) Initial stage (seven years eight months). (B) Active plate, which combined both upper and lower parts (seven years eleven months). (C) Active plate, which separated between upper and lower parts (nine years ten months). (D) Closing loop wire (11 years eleven months). (E) Postactive treatment stage (12 years nine months). (F) Postretention stage (26 years five months).

### Treatment progress

At the age of seven years eleven months, an active plate appliance that consisted of both upper and lower parts was applied to abolish the tongue thrust habit. The occlusal surface of the appliance was covered by resin to intrude both upper and lower molars (Figure 4B). This appliance was used for approximately two years. We focused attention particularly on oral hygiene because the enamel was thin. We also gave oral hygiene instructions during every patient visit.

After the tongue thrust habit was arrested, space closing of the upper and lower dentition was initiated using the same appliance (Figure 4C). At the age of

11 years eleven months, bands for the upper and lower first molars and lower first premolars were placed and an arch wire ( $0.017 \times 0.025$ " ) with a closing loop was applied. The spacing of the upper and lower anterior teeth closed within six months (Figure 4D), and the appliance was removed (Figures 2B, 3B, and 4E). A removable retainer was applied at the age of 12 years nine months. The retainer was removed at the age of 14 years one month because no relapse had been observed. At the age of 11 years, because of neuroparalysis, she was brought to our dental clinic in a wheelchair. Her physical condition had deteriorated further and she is bedridden because of the disease



**FIGURE 5.** Panoramic radiographs. (A) Initial stage (seven years eight months); (B) nine years four months; (C) twelve years five months; (D) postactive treatment stage (12 years nine months).

at present. After she was no longer able to come to our dental clinic, we visited the patient at home to check her occlusion and to perform oral hygiene instruction.

## RESULTS

The tongue thrust habit was completely eliminated using a removable appliance. Labial inclination of upper and lower anterior teeth was corrected and spacing of upper and lower dentition was closed completely (Figures 2B and 3B) and the protruded upper

lip was improved (Figures 1B,C and Figure 6). During the long-term retention period, optimal intercuspation of teeth was achieved and no space was observed, promoting good masticatory function (Figures 2C, 3C, and 4F). As a result, her occlusion was stable, and optimal intercuspation of the molar has been achieved at the age of 26 years five months (Figures 2C, 3C, and 4F; Table 1).

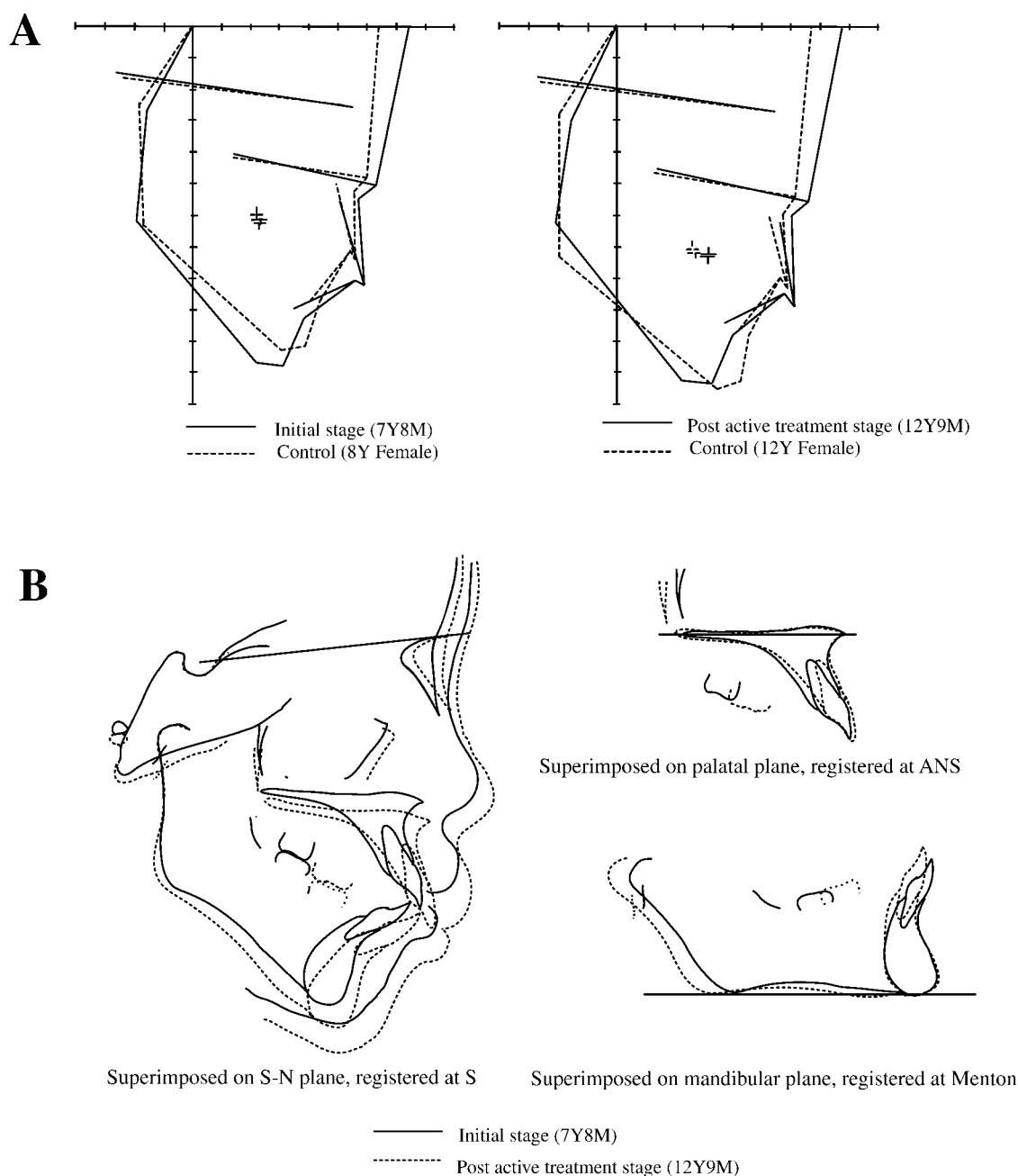
## DISCUSSION

Morquio syndrome is characterized by dwarfism, short neck, pigeon breast, spinal deformity, and knock knees.<sup>4</sup> The present case also showed these characteristics. There are few studies in which the craniofacial skeletal pattern of a patient with Morquio syndrome was examined using cephalometric radiographs. In the present case, the anteroposterior cranial length was larger than normal, but the anteroposterior relationship was skeletal Class I. The maxilla and mandible were located in a more posterior position, and the patient showed a high mandibular plane angle. This skeletal pattern has hardly changed during orthodontic treatment.

It was reported that a weak masticatory muscle contractile force might cause clockwise rotation of the mandible<sup>17</sup> and that a tongue thrust habit might cause labial inclination of anterior teeth.<sup>18</sup> It is known that patients with Morquio syndrome often show a weak muscle contraction force.<sup>4</sup> In the present case, the tongue thrust habit and weak masticatory muscle contraction force might have caused a labial inclination of the anterior teeth and clock-wise rotation of the mandible. Therefore, it was suggested that the prevention of the tongue thrust habit using an orthodontic appliance was possible, but correction of the skeletal pattern was considered difficult probably because of a weak masticatory muscle contraction force in such a patient.

Regarding the tooth quality of patients with Morquio syndrome, it was reported that such patients often have thin tooth enamel showing cracks and pits in the enamel.<sup>6-12</sup> The present case also showed these characteristics. Therefore, we provided strict oral hygiene instruction at each visit to prevent dental caries. Furthermore, we used a nonbonded orthodontic appliance instead of bonded orthodontic appliance such as edgewise appliance for the same reason. As a result, we completely prevented dental caries during the orthodontic treatment. Therefore, it is suggested that strict oral hygiene instruction and use of nonbonded orthodontic appliance are desirable in the orthodontic treatment of a patient with Morquio syndrome.

Regarding the occlusion in a patient with Morquio syndrome, it was reported that such a patient often showed labial inclination of the upper anterior teeth.<sup>18</sup>



**FIGURE 6.** Changes from initial to postactive treatment stage. (A) Profilogram. Left: initial stage (seven years eight months), solid line; control (eight years, girl), dashed line; right: postactive treatment stage (12 years nine months), solid line; control (12 years, girl), dashed line. (B) Superimposed tracings of lateral cephalograms. Solid line: initial stage (seven years eight months); dotted line: postactive treatment stage (12 years nine months).

The present case showed labial inclination of both upper and lower anterior teeth probably because of a tongue thrust habit.<sup>19</sup> In the present case, we used a removable orthodontic appliance similar to a Hawley retainer to arrest the habit and to intrude the molars of the maxilla and mandible because it was reported that such an appliance is effective in abolishing the tongue thrust habit. As a result, we abolished the tongue thrust habit completely.

However, the use of this removable appliance could not close the spaces in the upper and lower dentitions. Therefore, we used a fixed orthodontic appliance with closing loop and bands to close the spaces. As a result, we could close the spaces completely and the patient with Morquio syndrome was satisfied with the occlusion and masticatory function. Therefore, it is suggested that the use of a removable appliance and nonbonded fixed appliance with closing loop are de-

sirable to prevent the tongue thrust habit and to close spacing of anterior teeth in a patient with Morquio syndrome. We believe that such an orthodontic treatment contributed to increased QOL in such a patient.

## CONCLUSIONS

A patient with Morquio syndrome showing thin tooth enamel and malocclusion with spaced arches and labial inclination of the upper and lower anterior teeth probably because of a tongue thrust habit was provided strict oral hygiene instruction and treated with a removable orthodontic appliance and nonbonded fixed orthodontic appliance with closing loop. A stable good occlusion and good masticatory function were both achieved. However, correction of a skeletal abnormality was difficult probably because of a weak masticatory muscle contractile force.

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