

## Skeletal Characteristics and Treatment Outcome of Five Patients with Robin Sequence

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### ABSTRACT

**Objective:** To examine the variation in the precise skeletal characteristics and the treatment outcomes of five Japanese Robin sequence cases.

**Materials and Methods:** The birth histories and orthodontic records of five Japanese Robin sequence patients were collected and analyzed.

**Results:** All cases had a retrognathic appearance with small SNA and SNB angles. They had significantly steep mandibular planes with lingual tipped incisors in both arches. The gonial angles in two cases were within the Japanese norm, whereas the remaining three showed significantly enlarged angles. Moreover, all cases showed a significantly shorter ramus length, but the mandibular body was short in only two cases. All had moderate or severe crowding in both arches, and therefore extraction of lateral dentition or lateral incisors was performed in conjunction with orthodontic treatment. An edgewise multibracket appliance was placed, and labial tipping of the lower incisors was performed in all cases. All obtained normal functional occlusion after active treatment, but the retrognathic appearance remained in most cases.

**Conclusions:** The present cases with Robin sequence showed variation in the gonial angle and mandibular body length, although all commonly exhibited smaller SNA and SNB angles with significantly steep mandibular planes. Significant labial tipping of the lower incisors was required during the active treatment, and all cases finally obtained functional occlusion, indicating the relatively good prognosis on the occlusion of this sequence.

**KEY WORDS:** Robin sequence; Skeletal characteristics; Orthodontic treatment; Treatment outcome

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### INTRODUCTION

Robin sequence is characterized by retrognathic appearance, cleft palate, and glossoptosis.<sup>1</sup> The prevalence of Robin sequence has been reported to be 1/20,000 to 1/8500.<sup>2,3</sup> Patients commonly exhibit upper-airway obstruction and concomitant feeding difficulty.<sup>1</sup> Mandibular micrognathia or retrognathia is a primary pathogenetic event in which the tongue interferes with fusion of the palatal shelves and obstructs the upper airway.<sup>4,5</sup>

A number of cephalometric studies have reported the characteristic mandibular morphology of Robin sequence.<sup>6-14</sup> These reports consistently described the patients as showing a small mandible (especially ramus length) and steep mandibular plane; however, the size of the mandibular body and gonial angle varied among reports.<sup>8-14</sup> Regarding the outcome of orthodontic treatment, there have been only a few single case reports so far, limiting the useful information for treatment of this sequence.<sup>8,11</sup>

Therefore, in this study, we examined the precise

skeletal characteristics and morphological variation in the craniofacial region of Robin sequence patients and discussed the treatment outcome. To this end, five Japanese Robin sequence patients who came to our dental hospital were examined and compared both with the Japanese norm and with the mean values of isolated cleft palate patients.

## PATIENTS

A total of six Japanese cases were diagnosed as Robin sequence in our dental hospital. Among them, the skeletal characteristics and treatment outcome could not be evaluated in one case because the orthodontic treatment had just been initiated. Thus, the remaining five cases were shown and evaluated in this study. All the five cases were at IIIIB according to Hellman's dental age specifications.<sup>15</sup>

### Case 1

*Diagnosis and treatment objectives.* Case 1 was a male born to healthy parents and weighed 3760 g at birth. At the age of 2 months, he suffered from acute respiratory failure and underwent a tracheotomy. He had micrognathia and cleft palate and was diagnosed as Robin sequence. Palatoplasty was performed at 1 year 2 months of age.

He visited our dental hospital at 9 years 1 month of age and demonstrated a slight retrognathic appearance (Figure 1A). A deep bite with 6.0 mm of overbite was noted, and overjet was 3.5 mm. He was in the mixed dentition and had severe crowding (arch length discrepancy: maxilla, 11.4 mm; mandible, 10.5 mm). SNA and SNB angles were significantly smaller, and mandibular plane and gonial angles were much larger than the Japanese norm (Table 1).<sup>16,17</sup> The SNA-SNB difference was 5.4°. Both upper and lower incisors were significantly retroclined.

The treatment objectives were as follows: (1) to eliminate the arch length discrepancies in both arches by extraction and (2) to correct the deep bite and align both arches. Orthodontic treatment was initiated with a lingual arch to correct lingual displacement of the upper right and left lateral incisors. An edgewise multibracket appliance was placed in both arches with extraction of the upper right first premolar and left second premolar and lower first premolars at the age of 11 years 2 months.

*Treatment results.* Facial and oral photographs after the active treatment were taken at 15 years 10 months of age (Figure 1B). He obtained functional occlusion, but his retrognathic profile was somewhat worsened. Superimposed profilograms showed a large amount of vertical but a small amount of horizontal mandibular growth (Figure 1C).<sup>18</sup> The SNA angle did not change

but the SNB angle increased by 1.5° compared with the pretreatment values (Table 1). Both upper and lower incisors were significantly proclined during treatment.

### Case 2

*Diagnosis and treatment objectives.* Case 2 was a male born to healthy parents and weighed 2850 g at an uneventful birth. He had a cleft palate, and a palatoplasty was performed at 1 year 2 months of age.

He visited our dental hospital at 11 years 2 months of age with a slight retrognathic appearance (Figure 2A). A large overjet (7.0 mm) and a deep bite (overbite: 8.0 mm) were noted. Arch length discrepancies in the maxillary and mandibular arches were -6.0 and -11.3 mm, respectively. The lower left central and lateral incisors were fused. Both SNA and SNB were smaller than the Japanese norm<sup>16,17</sup> (Table 1). The SNB showed a significantly lower value, and the SNA-SNB difference was 8.0°. The gonial angle was near the norm, but the mandibular plane angle was significantly larger (Table 1). Both upper and lower incisors were significantly retroclined.

The treatment objectives were as follows: (1) to eliminate the arch length discrepancies in both arches by extraction and (2) to correct the deep bite and large overjet and align both arches. An edgewise multibracket appliance was placed in both arches, and the patient was treated with extraction of upper second premolars and the lower right canine and left first premolar.

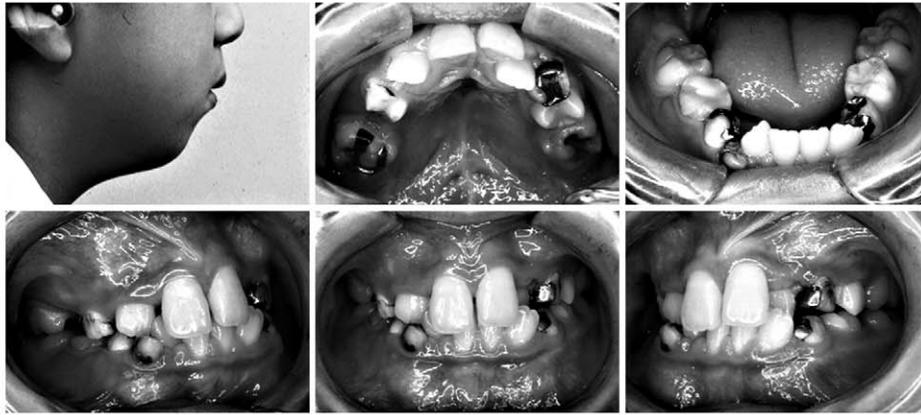
*Treatment results.* Facial and oral photographs after the active treatment were taken at 20 years 7 months of age (Figure 2B). The oral photographs showed a desirable occlusion, but the patient still had a slight retrognathic appearance. Downward and forward growth of the mandible had occurred (Figure 2C) and the SNB angle was increased by 2.0° (Table 1). In contrast, the SNA angle was unchanged compared with pretreatment. His upper incisors were retroclined and his lower incisors were significantly proclined during treatment.

### Case 3

*Diagnosis and treatment objectives.* Case 3 was a female born to healthy parents and weighed 2850 g at birth. At birth, she suffered from respiratory problems and therefore underwent oxygen inhalation. She had cleft palate and retrognathic appearance and was diagnosed as Robin sequence. Palatoplasty was performed at 1 year 2 months of age.

She visited our dental hospital at 9 years 1 month of age. A facial photograph taken at this time showed an apparent retrognathic appearance (Figure 3A). A large overjet (7.0 mm) and a deep bite (overbite: 7.0

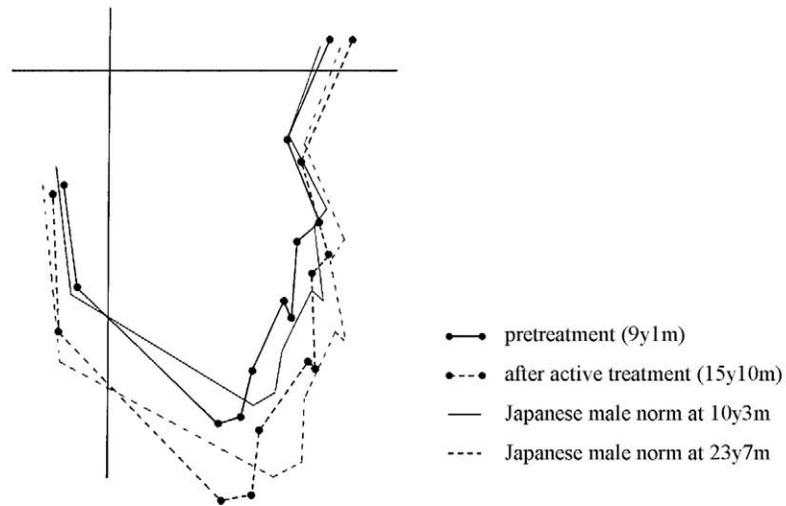
**A** Facial and oral photographs at pretreatment



**B** Facial and oral photographs at after active orthodontic treatment



**C** Superimposed profilograms



**Figure 1.** Case 1. Facial and oral photographs. (A) Pretreatment. (B) After active orthodontic treatment. (C) Superimposed profilograms.<sup>18</sup>

**Table 1.** Cephalometric Measures Before and After the Active Treatment<sup>a</sup>

	Case 1		Case 2		Case 3		Case 4		Case 5	
	Pretreatment (9 y 1 mo)	Posttreatment (15 y 10 mo)	Pretreatment (11 y 2 mo)	Posttreatment (20 y 7 mo)	Pretreatment (9 y 1 mo)	Posttreatment (16 y 2 mo)	Pretreatment (10 y 7 mo)	Posttreatment (19 y 10 mo)	Pretreatment (9 y 3 mo)	Posttreatment (16 y 3 mo)
S-N	63.1 (61.0 ± 2.6)	68.0 (65.3 ± 2.4)	66.0 (62.2 ± 2.6)	69.0 (65.3 ± 2.4)	62.0 (60.3 ± 2.4)	63.0 (63.1 ± 3.0)	56.0 (60.9 ± 2.3)	56.5 (63.1 ± 3.0)	62.3 (60.3 ± 2.4)	65.9 (63.1 ± 3.0)
S-Ba	52.3 (50.5 ± 2.5)	56.9 (57.6 ± 3.1)	49.7 (52.2 ± 2.5)	57.8 (57.6 ± 3.1)	44.7 (49.6 ± 2.0)	49.4 (53.9 ± 2.4)	46.7 (50.5 ± 2.0)	48.0 (53.9 ± 2.4)	45.0 (49.6 ± 2.0)	50.0 (53.9 ± 2.4)
SNA	72.4 (80.9 ± 3.1)	73.0 (81.8 ± 3.1)	75.5 (80.9 ± 3.1)	75.5 (81.8 ± 3.1)	72.5 (80.9 ± 3.1)	72.5 (82.3 ± 3.5)	76.0 (80.9 ± 3.1)	76.0 (82.3 ± 3.5)	75.5 (80.9 ± 3.1)	75.5 (82.3 ± 3.5)
ANS-PNS	47.7 (48.8 ± 2.2)	52.4 (54.1 ± 2.5)	50.5 (50.2 ± 2.1)	56.5 (54.1 ± 2.5)	44.5 (49.5 ± 2.5)	53.4 (53.3 ± 2.5)	42.3 (50.1 ± 2.5)	44.1 (53.3 ± 2.5)	46.7 (49.5 ± 2.5)	50.2 (53.3 ± 2.5)
U-1 to FH plane	97.0 (109.8 ± 5.3)	112.2 (108.9 ± 5.6)	101.0 (109.8 ± 5.3)	90.0 (108.9 ± 5.6)	101.0 (109.8 ± 5.3)	105.6 (111.1 ± 5.5)	98.5 (109.8 ± 5.3)	107.4 (111.1 ± 5.5)	107.2 (109.8 ± 5.3)	90.0 (111.1 ± 5.5)
SNB	67.0 (76.2 ± 2.8)	68.5 (78.6 ± 3.1)	67.5 (76.2 ± 2.8)	69.5 (78.6 ± 3.1)	63.6 (76.2 ± 2.8)	64.6 (78.9 ± 3.5)	73.0 (76.2 ± 2.8)	73.0 (78.9 ± 3.5)	68.0 (76.2 ± 2.8)	69.2 (78.9 ± 3.5)
SNA-SNB diff.	5.4 (4.8)	4.5 (3.3 ± 2.7)	8.0 (4.8)	6.0 (3.3 ± 2.7)	8.9 (4.8)	7.9 (3.4 ± 1.8)	3.0 (4.8)	3.0 (3.4 ± 1.8)	7.5 (4.8)	6.3 (3.4 ± 1.8)
Mandibular plane angle	45.3 (32.0 ± 2.4)	45.0 (26.3 ± 6.3)	45.4 (32.0 ± 2.4)	42.5 (26.3 ± 6.3)	44.7 (32.0 ± 2.4)	47.7 (28.8 ± 5.2)	43.0 (32.0 ± 2.4)	43.5 (28.8 ± 5.2)	42.5 (32.0 ± 2.4)	43.0 (28.8 ± 5.2)
Gonial angle	141.0 (129.2 ± 4.7)	140.3 (119.4 ± 5.8)	131.0 (129.2 ± 4.7)	129.0 (119.4 ± 5.8)	144.3 (129.2 ± 4.7)	145.7 (122.2 ± 4.6)	133.3 (129.2 ± 4.7)	130.0 (122.2 ± 4.6)	138.6 (129.2 ± 4.7)	131.6 (122.2 ± 4.6)
Ramus incli- nation	5.8 (82.6 ± 5.6)	5.1 (2.6 ± 4.1)	-4.3 (82.6 ± 5.6)	-3.2 (2.6 ± 4.1)	9.6 (82.6 ± 5.6)	8.0 (2.9 ± 4.4)	-0.3 (82.6 ± 5.6)	-3.6 (2.9 ± 4.4)	8.4 (82.6 ± 5.6)	3.9 (2.9 ± 4.4)
Ar-Go	32.2 (41.4 ± 3.9)	41.2 (49.4 ± 5.2)	37.4 (43.0 ± 4.3)	45.3 (49.4 ± 5.2)	33.7 (41.6 ± 3.3)	37.5 (47.7 ± 4.4)	37.8 (42.6 ± 3.2)	41.0 (47.7 ± 4.4)	37.4 (41.6 ± 3.3)	42.0 (47.7 ± 4.4)
Go-Pog	64.3 (67.4 ± 3.6)	73.3 (77.3 ± 4.3)	69.9 (70.3 ± 3.5)	76.3 (77.3 ± 4.3)	58.3 (66.8 ± 2.4)	68.8 (74.9 ± 3.7)	62.1 (68.6 ± 2.0)	68.9 (74.9 ± 3.7)	67.5 (66.8 ± 2.4)	79.0 (74.9 ± 3.7)
Ar-Pog	91.2 (100.2)	107.3 (116.0)	97.9 (104.2)	110.0 (116.0)	87.2 (99.6)	101.4 (111.2)	91.3 (102.0)	99.7 (111.2)	98.0 (99.6)	111.0 (111.2)
L-1 to Man- dibular plane	79.5 (93.8 ± 5.9)	88.5 (94.7 ± 7.2)	85.0 (93.8 ± 5.9)	98.0 (94.7 ± 7.2)	75.7 (93.8 ± 5.9)	96.5 (96.3 ± 5.8)	65.2 (93.8 ± 5.9)	83.3 (96.3 ± 5.8)	77.5 (93.8 ± 5.9)	91.3 (96.3 ± 5.8)

<sup>a</sup> S indicates sella turcica; N, nasion; A, point A; SNA, angle between SN and NA; U-1, long axis of maxillary central incisor; U-1 to FH plane, angle between U-1 and FH plane; B, point B; SNB, angle between SN and NB; Mandibular plane angle, angle between mandibular plane and frankfort horizontal (FH) plane; Gonial angle, angle between mandibular plane and ramus plane; Ramus inclination, angle between ramus plane and FH plane; Ar, articulare; Go, gonion; Ar-Go, distance between Ar and Go; Pog, pogonion; Go-Pog, distance between Go and Pog; L-1, long axis of mandibular central incisor; L-1 to mandibular plane, angle between L-1 and mandibular plane. Sex and age-matched Japanese norm ± SD are in parentheses.<sup>16,17</sup>

mm) were noted. The lower right lateral incisor was congenitally absent and the upper left lateral incisor was a microtooth. Arch length discrepancies in the maxilla and mandible were -11.4 and -7.6 mm, respectively. Both the SNA and SNB angles were significantly smaller and the mandibular plane and gonial angles were much larger than the Japanese norm.<sup>16,17</sup> The SNA-SNB difference was 8.9° (Table 1). Both upper and lower incisors were significantly retroclined.

The treatment objectives were as follows: (1) to eliminate the arch length discrepancies in both arches by extraction and (2) to correct the deep bite and large overjet and align both arches. An edgewise multi-bracket appliance was placed in both arches with extraction of the upper left lateral incisor and second premolar.

**Treatment results.** Facial and oral photographs after the active treatment were taken at 16 years 2 months of age (Figure 3B). Her retrognathic profile was improved, and she obtained functional occlusion. The mandibular growth was mainly toward the downward direction (Figure 3C). The SNA angle was unchanged, but the SNB angle increased by 1° (Table 1). Signifi-

cant labial tipping of the lower incisors was noted (Table 1).

**Case 4**

**Diagnosis and treatment objectives.** Case 4 was a female born to healthy parents and weighed 2320 g at birth. She had micrognathia and cleft palate. At the age of 3 months, she had cyanosis and was diagnosed as Robin sequence. Palatoplasty was performed at 2 years of age.

She visited our dental hospital at 10 years 7 months of age with a slight retrognathic appearance (Figure 4A). Deep bite was noted with 8.0-mm overbite and overjet of 2.0 mm. Arch length discrepancies in the maxilla and mandible were -10.6 and -4.9 mm, respectively. The lower right lateral incisor was congenitally absent, and the upper right and left lateral incisors were microteeth. SNA and SNB angles were smaller than the Japanese norm, and SNA-SNB difference was near the Japanese norm (Table 1).<sup>16,17</sup> The mandibular plane angle was larger, but the gonial angle was not significantly different from the Japanese

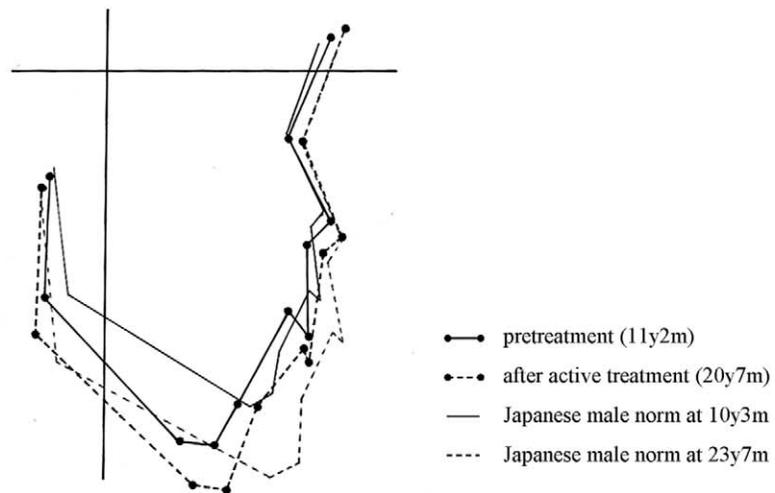
**A** Facial and oral photographs at pretreatment



**B** Facial and oral photographs at after active orthodontic treatment

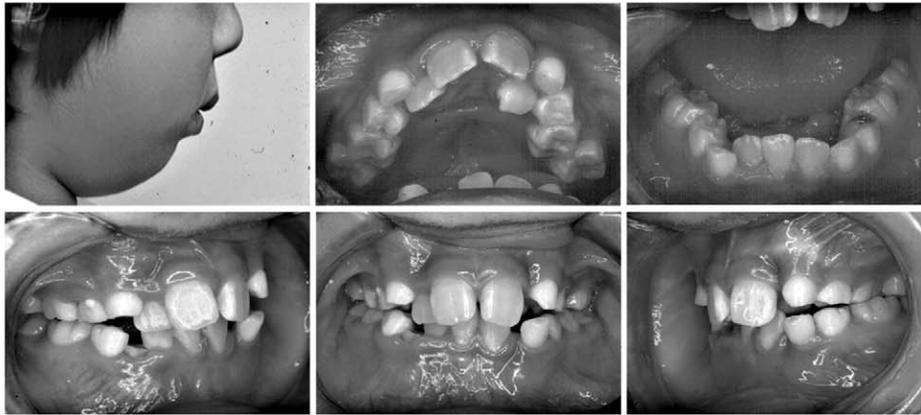


**C** Superimposed profilograms



**Figure 2.** Case 2. Facial and oral photographs. (A) Pretreatment. (B) After active orthodontic treatment. (C) Superimposed profilograms.<sup>18</sup>

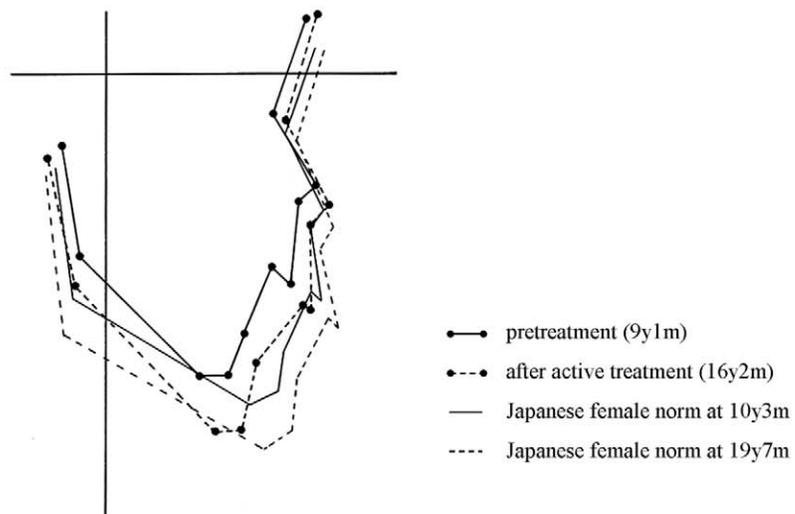
**A** Facial and oral photographs at pretreatment



**B** Facial and oral photographs at after active orthodontic treatment

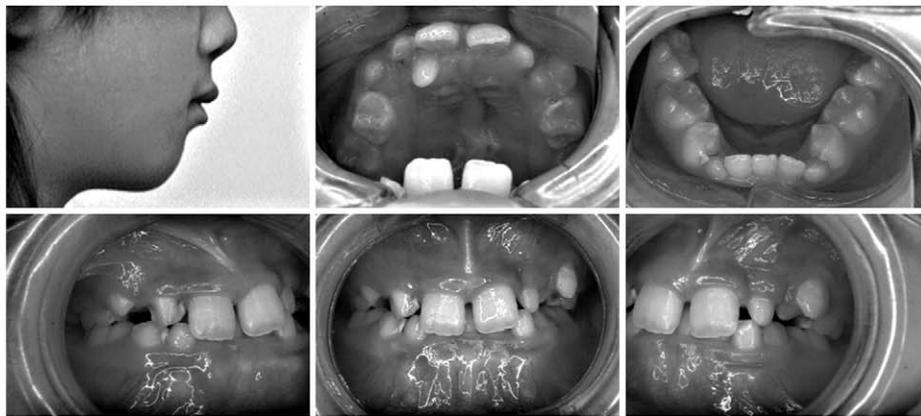


**C** Superimposed profilograms



**Figure 3.** Case 3. Facial and oral photographs. (A) Pretreatment. (B) After active orthodontic treatment. (C) Superimposed profilograms.<sup>18</sup>

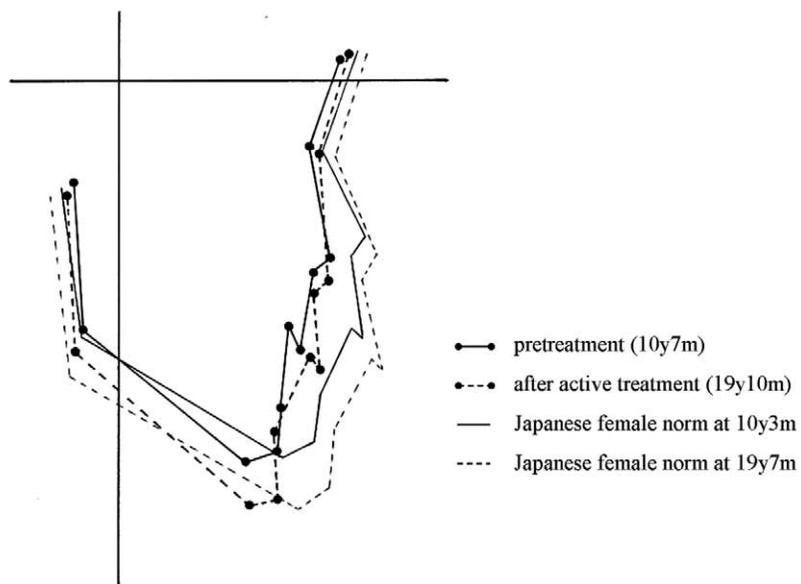
**A** Facial and oral photographs at pretreatment



**B** Facial and oral photographs at after active orthodontic treatment



**C** Superimposed profilograms



**Figure 4.** Case 4. Facial and oral photographs. (A) Pretreatment. (B) After active orthodontic treatment. (C) Superimposed profilograms.<sup>18</sup>

norm.<sup>16</sup> Both upper and lower incisors were significantly retroclined.

The treatment objectives were as follows: (1) to expand the maxillary arch laterally, (2) to promote the mandibular growth, (3) to eliminate the arch length discrepancies in both arches, and (4) to correct the deep bite and align both arches. Orthodontic treatment was initiated with a rapid palatal expansion appliance or expander, followed by the use of a jumping plate to promote mandibular growth. At 15 years of age, an edgewise multibracket appliance was placed in both the arches with extraction of upper right and left lateral incisors.

**Treatment results.** Facial and oral photographs after the active treatment were taken at 19 years 10 months of age (Figure 4B). A facial photograph taken at this time still showed a slight retrognathic profile, but functional occlusion was attained. Prosthetic treatment was planned in the position of the upper left lateral incisor. Growth of the mandible was entirely in a downward direction (Figure 4C). There was no increase in the SNA and SNB angles (Table 1). Significant labial tipping of the upper and lower incisors was noted (Table 1).

### Case 5

**Diagnosis and treatment objectives.** Case 5 was a female born to healthy parents and weighed 3020 g at birth. She exhibited characteristic micrognathia and cleft palate at birth and was diagnosed as Robin sequence. Palatoplasty was performed at 1 year 2 months of age.

She visited our dental hospital at 9 years 3 months of age. A retrognathic appearance was noted from a facial photograph (Figure 5A). A large overjet of 10.0 mm and an overbite of 4.0 mm were seen. Severe crowding was seen in both arches (arch length discrepancy: maxilla, 12.2 mm; mandible, 9.4 mm). Both SNA and SNB angles were smaller than the Japanese norm<sup>16,17</sup> (Table 1). The SNB showed a significantly lower value and the SNA-SNB difference was 7.5° (Table 1). The mandibular plane and gonial angles were much larger than the Japanese norm. The lower incisors were significantly retroclined, but the inclination of the upper incisors was almost the same as the Japanese norm.

The treatment objectives were as follows: (1) to expand the maxillary arch laterally, (2) to eliminate the arch length discrepancies in both arches, and (3) to correct the large overjet and align both arches. Orthodontic treatment was initiated with a Porter-type maxillary expansion appliance, followed by treatment with an edgewise multibracket appliance in both the arches with extraction of upper and lower first premolars.

**Treatment results.** Facial and oral photographs after

the active treatment were taken at 16 years 3 months of age, and good functional occlusion was obtained (Figure 5B). However, she still exhibited a retrognathic profile. Downward and forward growth of the mandible was seen (Figure 5C). The SNA angle did not change, but the SNB angle increased by 1.2° during treatment (Table 1). Significant lingual and labial tipping of upper and lower incisors was noted, respectively (Figure 5C).

### Pretreatment Skeletal Characteristics

To evaluate the skeletal characteristics of the five patients, cephalometric measurements of the cranial base, maxilla, and mandible were examined and compared with the Japanese norm<sup>16,17</sup> at IIIB according to Hellman's dental age specifications,<sup>15</sup> as shown in Figure 6. The mean values of orthodontically untreated isolated cleft palate patients (CP) are also included in this figure. These mean values were generated from 28 nonsyndromic Japanese CP (9 males and 19 females, from 8 years to 11 years and 11 months of age) at IIIB according to Hellman's dental age specifications.<sup>15</sup>

The dentofacial morphology of the five presented Robin sequence patients was characterized by a smaller SNB angle and a larger mandibular plane angle than the Japanese norm<sup>16,17</sup> and the mean values of CP patients (Figure 6). A significantly shorter Ar-Go length was also noted. The upper and lower incisors were more retroclined than the Japanese norm and the mean values of CP. The mean values of gonial angle and Go-Pog length of the presented five cases were larger and shorter than the mean values of CP, respectively. However, in some cases, they were not largely different from the Japanese norm.<sup>16,17</sup>

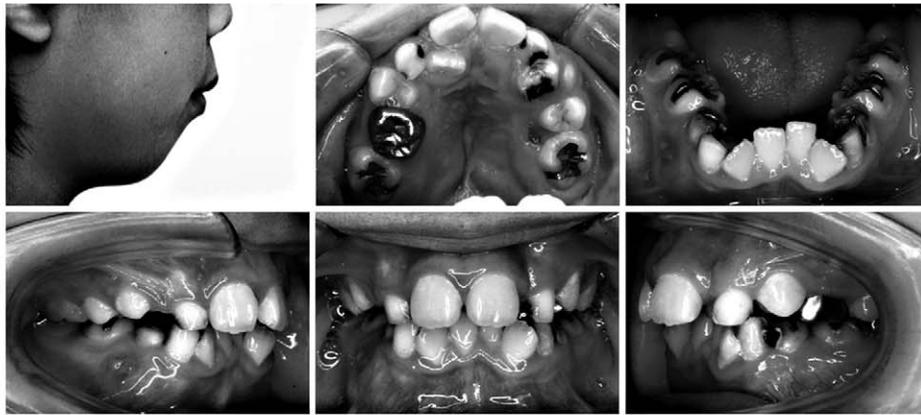
### Summary of Treatment Outcome of Five Presented Cases

All cases, except case 4, showed a decreased ANB angle with a slightly increased SNB angle (Table 1). The change in the mandibular plane varied among cases. It increased in case 3 but decreased in case 2 and remained almost unchanged in cases 1, 4, and 5. The upper incisor was proclined in cases 1, 3, and 4 and retroclined in cases 2 and 5. In contrast, the lower incisor was significantly proclined in all cases.

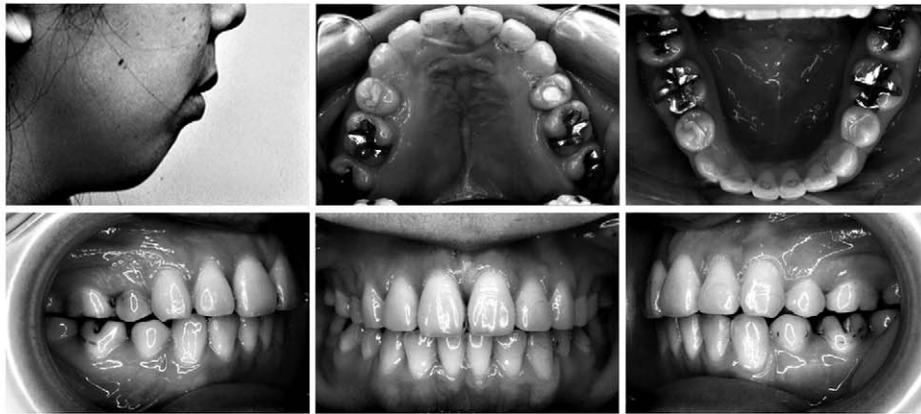
### Mandibular Growth of Five Presented Cases

Because a slight increase in the SNB angle was seen in cases 1, 2, 3, and 5, the increments in Ar-Go, Go-Pog, and Ar-Pog were calculated and compared with the Japanese norm<sup>17</sup> (Table 2). The increase in Ar-Go was above the norm in cases 1, 4, and 5 and Go-Pog was above the norm in cases 3 and 5. The

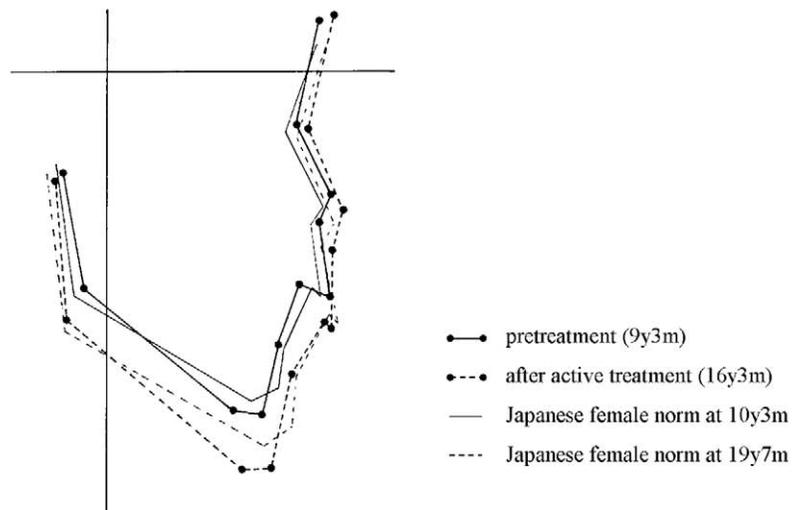
**A** Facial and oral photographs at pretreatment



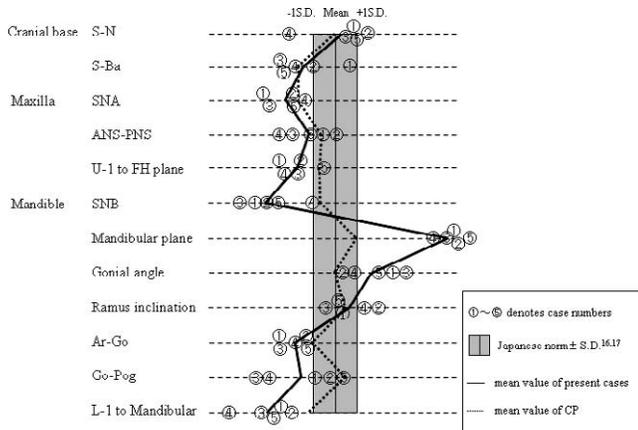
**B** Facial and oral photographs at after active orthodontic treatment



**C** Superimposed profilograms



**Figure 5.** Case 5. Facial and oral photographs. (A) Pretreatment. (B) After active orthodontic treatment. (C) Superimposed profilograms.<sup>18</sup>



**Figure 6.** Pretreatment skeletal characteristics of the presented five Robin sequence cases with the Japanese norm<sup>16,17</sup> and the mean values of untreated isolated cleft palate patient (CP). Each value is represented by Z score [(measurement – Japanese norm)/SD].

increase in Ar-Pog was above the norm in cases 1, 2, 3, and 5.

**Tooth Abnormalities**

Of the five presented cases, three showed tooth abnormalities in the lower incisors (Table 3). Case 2 had fusion of the lower left central and lateral incisors. Case 3 had a congenitally absent lower right lateral incisor and the upper left lateral incisor was a microtooth. Case 4 had a congenitally absent lower right lateral incisor and the upper right and left lateral incisors were microteeth.

**DISCUSSION**

The five presented Japanese cases of Robin sequence commonly exhibited a small SNB angle, a

steep mandibular plane, and a short ramus length (Figure 6; Table 1), as reported previously.<sup>8-14</sup> The gonial angle in cases 1, 3, and 5 was significantly higher than the Japanese norm<sup>16,17</sup> and the mean values of CP patients (Figure 6; Table 1), but this was not the case in cases 2 and 4. Moreover, the mandibular body length (Go-Pog) was significantly shorter in cases 3 and 4 but not in cases 1, 2, and 5. These findings clearly support the variation of the mandibular morphology reported in previous studies<sup>8-14</sup> and demonstrate that both micrognathia and retrognathia are seen in Robin sequence.<sup>4</sup> The exact reason for this variation is not known but could be related to the diversity of the etiology of this sequence.<sup>4,5,19,20</sup> The size of the airway and the tongue position, which could not be examined in the present sample, might be also related to this variation.

A high rate of tooth abnormality (especially congenitally absent teeth) was seen in the lower arch of the presented cases (Table 3). This is consistent with the previous report that congenitally absent teeth were twice as common in the lower arch than in the upper.<sup>21</sup> However, all the absent teeth were lower incisors in the present five cases, unlike the study reporting that the absent teeth were most frequently seen in the lower second premolars.<sup>21</sup>

In addition to the SNB angle, the SNA angle and ANS-PNS length of all presented cases were significantly smaller than the Japanese norm (Figure 6; Table 1). The mean value of these cases was even smaller than that of CP patients (Figure 6). This finding is consistent with a previous report in which maxillary anterior-posterior length was significantly shorter in Robin sequence patients than CP.<sup>12</sup> The exact reason of this shortness is not known. However, deformation of the mandible be-

**Table 2.** Mandibular Growth in Present Five Cases (%)<sup>a,b</sup>

	Active Treatment Period				
	Case 1 (9 y 1 mo– 15 y 10 mo)	Case 2 (11 y 2 mo– 15 y 2 mo)	Case 3 (9 y 1 mo– 14 y 8 mo)	Case 4 (10 y 7 mo– 14 y 4 mo)	Case 5 (9 y 3 mo– 14 y 10 mo)
Ar-Go	113.5	88.1	82.7	103.9	129.7
Go-Pog	91.2	95.0	110.4	96.1	128.2
Ar-Pog	101.3	100.8	127.9	95.4	116.2

<sup>a</sup> Ar indicates articulare; Go, gonion; Ar-Go, distance between Ar and Go; Pog, pogonion; Go-Pog, distance between Go and Pog; Ar-Pog, distance between Ar and Pog.

<sup>b</sup> Each value is represented as the percentage of the calculated Japanese norm.<sup>17</sup>

**Table 3.** Tooth Abnormality of Present Five Cases

	Case 1	Case 2	Case 3	Case 4	Case 5
Congenitally missing teeth	Not seen	Not seen	Mandibular lateral incisor	Mandibular lateral incisor	Not seen
Malformed teeth	Not seen	Mandibular incisor (fusion tooth)	Maxillary lateral incisor (microtooth)	Maxillary lateral incisors (microteeth)	Not seen

cause of fetal head constraint is thought to be one of the etiological factors of this sequence.<sup>4,5,19,20</sup> Head constraint is known to cause deformation in the cranial region.<sup>22</sup> Thus, it is no wonder that the maxilla was affected by the constraint, resulting in the maxillary growth disturbance in the present cases.

In Robin sequence, a larger increment of mandibular growth, the so-called catch-up growth, is frequently seen.<sup>9</sup> In contrast, Daskalogiannakis et al reported that this type of growth could not be seen after 5 years of age.<sup>12</sup> In this study, the increment in Go-Pog was larger than the Japanese norm in cases 3 and 5 (Table 2) and SNB angle increased in cases 1, 2, 3, and 5 during the active treatment (Table 1). However, because all five cases presented in this study underwent orthodontic treatment, it is difficult to clear up this controversial issue on the natural growth by examining present cases. Despite the observed mandibular growth, proclination of the lower incisors was required to obtain good functional occlusion in all cases. This was because they uniformly exhibited a steep mandibular plane and greater mandibular growth in the vertical dimension.

Unfortunately, except in case 3, the retrognathic appearance was not improved after the treatment (Figures 1B through 5B). As a treatment alternative, genioplasty could be carried out, and this would be the most reliable treatment plan to improve the profile, provided the patients are not concerned about surgical intervention or admission to a hospital.

## REFERENCES

- Randall P. Micrognathia and glossoptosis with airway obstruction: the Pierre Robin syndrome. In: Converse JM, ed. *Reconstructive Plastic Surgery*. Philadelphia, Penn: WB Saunders; 1964:1452–1462.
- Bush PJ, Williams AJ. Incidence of the Robin Anomalad (Pierre Robin syndrome). *Br J Plast Surg*. 1983;36:434–437.
- Tolarova MM, Cervenka J. Classification and birth prevalence of orofacial clefts. *Am J Med Genet*. 1998;75:126–137.
- Cohen MM Jr. Robin sequences and complexes: causal heterogeneity and pathogenetic/phenotypic variability. *Am J Med Genet*. 1999;84:311–315.
- Cohen MM Jr. Interface between Robin sequence and ordinary cleft palate. *Am J Med Genet*. 2001;101:288.
- Pruzansky S. Not all dwarfed mandibles are alike. *Birth Defects*. 1969;5:120–129.
- Hotz M, Gnoinski W. Clefts of the secondary palate associated with the “Pierre Robin syndrome.” *Swed Dent J*. 1982;6(suppl):89–98.
- Koubayashi S, Takata Y, Susami R, Morikawa M, Sakuda M. A case of Robin anomalad in association with orthodontic treatment. *J Jpn Orthod Soc*. 1986;45:295–310.
- Figueroa AA, Glupker TJ, Fitz MG, BeGole EA. Mandible, tongue, and airway in Pierre Robin sequence: a longitudinal cephalometric study. *Cleft Palate Craniofac J*. 1991;28:425–434.
- Laitinen SH, Ranta RE. Cephalometric measurements in patients with Pierre Robin syndrome and isolated cleft palate. *Scand J Plast Reconstr Surg Hand Surg*. 1992;26:177–183.
- Okada K, Yamashiro T, Tenshin S, Takano-Yamamoto T. Orthodontic treatment for a patient with Pierre-Robin sequence complicated by juvenile periodontitis. *Cleft Palate Craniofac J*. 2000;37:318–324.
- Daskalogiannakis J, Ross RB, Tompson BD. The mandibular catch-up growth controversy in Pierre Robin sequence. *Am J Orthod Dentofacial Orthop*. 2001;120:280–285.
- Hermann NV, Kreiborg S, Darvann TA, Jensen BL, Dahl E, Bolund S. Early craniofacial morphology and growth in children with nonsyndromic Robin Sequence. *Cleft Palate Craniofac J*. 2003;40:131–143.
- Hermann NV, Kreiborg S, Darvann TA, Jensen BL, Dahl E, Bolund S. Craniofacial morphology and growth comparisons in children with Robin Sequence, isolated cleft palate, and unilateral complete cleft lip and palate. *Cleft Palate Craniofac J*. 2003;40:373–396.
- Hellman M. Development of face and dentition in its application to orthodontic treatment. *Am J Orthod*. 1940;26:424–447.
- Iizuka T. Roentgencephalometric analysis of craniofacial growth in Japanese children. *J Stomatol Soc Jpn*. 1958;25:18–30.
- Masaki F. The longitudinal study of morphological differences in the cranial base and facial structure between Japanese and American white. *J Jpn Orthod Soc*. 1980;39:436–456.
- Sakamoto T. A study on the developmental changes of dentofacial complex of the Japanese with special reference to sella turcica. *J Jpn Orthod Soc*. 1959;18:1–17.
- Latham RA. The pathogenesis of cleft palate associated with the Pierre Robin syndrome. An analysis of a seventeen-week human foetus. *Br J Plast Surg*. 1966;19:205–214.
- Szawillo D. The Pierre Robin syndrome: etiology and early treatment. *Trans Int Conf Oral Surg*. 1967:425–429.
- Ranta R, Rintala AE. The Pierre robin anomaly-comparisons of some disturbances in the formation of the teeth and the lower lip. *Proc Finn Dent Soc*. 1983;79:155–161.
- Graham JM Jr, Badura RJ, Smith DW. Coronal craniostenosis: fetal head constraint as one possible cause. *Pediatrics*. 1980;65:995–999.

## APPENDIX

In addition to 28 non-syndromic Japanese CP at IIIB according to Hellman's dental age specifications<sup>15</sup> (9 males and 19 females, aged from 8 years to 11 years and 11 months old), 19 cases of non-syndromic Japanese CP (not at IIIB) were added to the sample. Congenitally absent teeth (except third molars in both arches) were examined in total 47 cases (12 males and 35 females). 20 cases out of 47 cases had congenitally absent teeth (42.6%). The missing teeth were 38.7% upper incisors, 41.9% upper premolars, 12.9% lower incisors and 6.5% premolars.