Case Report

Long-Term Changes in Dentoskeletal Pattern in a Case with Beckwith-Wiedemann Syndrome Following Tongue Reduction and Orthodontic Treatment

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Abstract: Long-term changes in the dentoskeletal pattern in a 6-year-old Japanese girl with Beckwith-Wiedemann syndrome were demonstrated. The patient showed macroglossia, which is the most common symptom of the syndrome, protruded lower lip, mandibular protrusion and anterior open bite. The jaw base relationship improved to skeletal Class I and the molar relationship to Angle Class I at the early preadolescent period following tongue reduction and phase I orthodontic treatment using a chin cap and tongue crib. Optimum intercuspation of teeth was achieved after edgewise treatment without orthognathic surgery, and a skeletal Class I apical base relationship and good facial profile were maintained after the retention period of 2 years. This case report suggests that early orthodontic treatment with tongue reduction can be effective in a case with Beckwith-Wiedemann syndrome to improve an abnormal dentoskeletal pattern. (*Angle Orthod* 2000;70:326–331.)

Key Words: Beckwith-Wiedemann syndrome; Macroglossia; Tongue reduction; Orthodontic treatment

INTRODUCTION

Beckwith in 1963 and Wiedemann in 1964 originally reported Beckwith-Wiedemann syndrome. Macroglossia is the predominent finding in Beckwith-Wiedemann syndrome (97%); however, other findings include postnatal somatic gigantism (88%), abdominal wall defects and hernias (80%), abnormal earlobe creases/pits (76%), hypoglycemia (63%), nevus flammeus of the face (62%), nephromegaly (59%), hemihypertrophy (24%), congenital heart defects (6.5%) and cleft palate (2.5%).[1–4] Neurologic problems or mental retardation occurs in only a minority of patients, and is likely the result of hypoglycemic attacks in the neonatal period. Childhood neoplasms develop in some of the people affected by this syndrome and most are malignant. They include nephroblastoma, adrenocortical cancer, and hepatoblastoma. Some experts feel that all of these patients should be screened for cancer with regular abdominal ultrasound and serum alfa-fetoprotein levels.

Previous studies have shown that one major effect of macroglossia is a protrusion of dentoalveolar structures, which results in a protruding mandible, anterior open bite, abnormally obtuse gonial angle, and increased mandibular length.[5–7] Therefore, early intervention by tongue reduction,[8] early functional treatment of the stomatognathic system,[9] or both, is recommended in order to prevent both mandibular prognathism and anterior open bite.

There have been few case reports or studies concerning the longitudinal dentoskeletal changes in patients with Beckwith-Wiedemann syndrome treated with tongue reduction and orthodontic treatment. This article demonstrates the successful treatment and the long-term changes in dentoskeletal pattern in a patient with Beckwith-Wiedemann syndrome who was treated by tongue reduction and phase I and II orthodontic treatments.

CASE REPORT

Preadolescence

A 6-year-old Japanese girl with macroglossia, mandibular protrusion, and anterior open bite was diagnosed with Beckwith-Wiedemann syndrome characterized by macroglossia, postnatal somatic gigantism, and nephromegaly at a national hospital in Japan. The pediatrician at the hospital

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Accepted: March 2000. Submitted: March 2000.

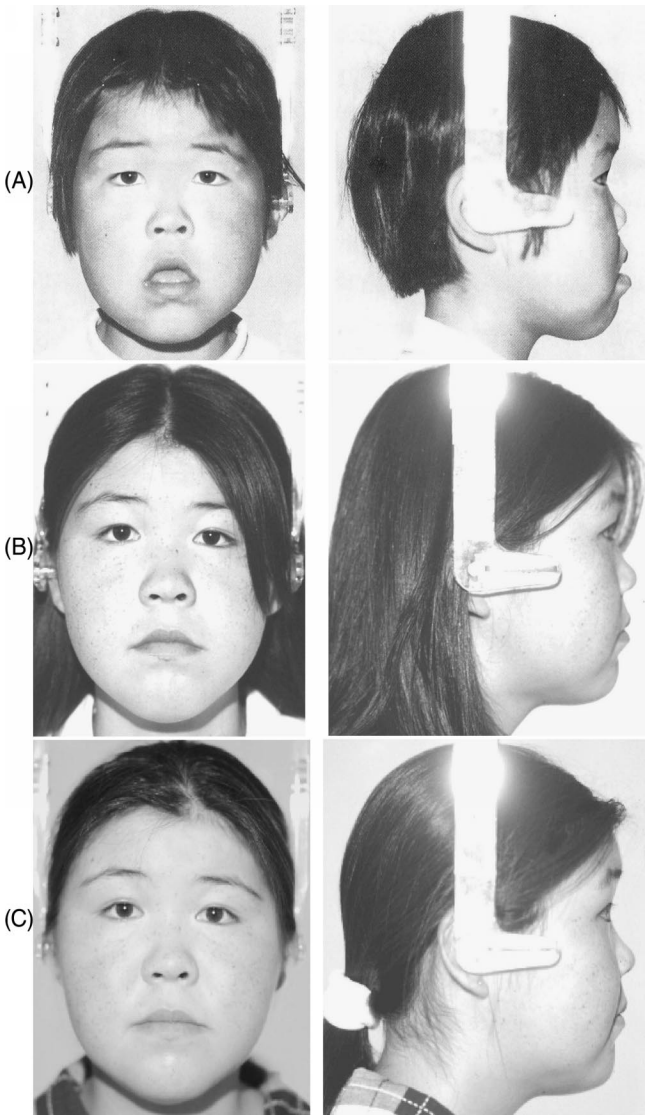

FIGURE 1. Facial photographs. (A) Initial stage (6Y4M); (B) Post-active-treatment stage (15Y8M); (C) Post-retention stage (18Y7M); Left: Frontal view; Right: Lateral view.

detected dentoskeletal disharmony, and orthognathic treatment was initiated at a University Dental Hospital. Family history revealed no notable hereditary diseases.

Clinical examination. The patient had a concave profile and a severely protruded lower lip (Figure 1). The tongue was large and, when extended, was long enough to reach the mentum (Figure 2). Mandibular protrusion and an anterior open bite were present. The molar relationship was Angle Class III, and all teeth showed cross bite relationships (Figure 3). Lateral cephalometrics showed a skeletal Class III apical base relationship, normal-sized maxilla, anterior displacement of the mandible, long mandibular length, and obtuse gonial angle when compared with the normative values for Japanese girls of corresponding age (Figure 4; Table 1).[10] Such a condition hindered verbal abilities and the patient experienced difficulty in pronouncing ''s,'' ''t,'' and ''l'' sounds.

Treatment plan. Treatment was planned as follows: (1) tongue reduction by the Becker method,[11] (2) inhibition of mandibular growth and tongue protrusion using a chin cap and tongue crib, and (3) review the diagnosis after completion of the permanent dentition.

Treatment progress. Tongue reduction was performed at 8 years of age using the Becker method.[11] A nighttime chin cap appliance was begun 2 months after the surgery and continued for 1 year and 5 months. A tongue crib was used from age 8 years 7 months old for 7 months.

Results achieved. The protruded lower lip was corrected (Figure 1) and the tongue volume was reduced (Figure 2). The skeletal Class III jaw base relationship and anterior open bite improved rapidly after tongue reduction and phase I orthodontic treatment (Figures 4 and 5). Total cross bite was partially corrected, and Angle Class I molar relationship was achieved, but a slight anterior open bite remained (Figure 5). The difficulties in pronunciation found prior to the tongue reduction were improved.

Adolescence

The dentoskeletal disharmony was corrected by tongue reduction and phase I orthodontic treatment and the patient was observed for 2 years and 4 months. At the end of this observation period (11 years 6 months of age) data comparable to the initial stage were recorded. At this time the profile was straight and moderate crowding with slight anterior open bite was observed (Figure 5). Lateral cephalometric radiographs showed a skeletal Class I jaw base relationship and average mandibular plane angle (Figure 4; Table 1) when compared to the normative values for Japanese girls of the corresponding age.[10] The molar relationship was Angle Class I (Figure 5).

Treatment plan. Treatment was planned as follows: (1) alignment of teeth using an edgewise appliance, and (2) retention with a Hawley-type retainer and fixed lingual retainer on the upper and lower dentitions, respectively.

Treatment progress. Edgewise treatment was initiated at age 11 years 7 months of age. Space closure was performed for 1 year after leveling. Up and down elastics were used for 6 months and detailing was conducted for 1 and a half years. The appliance was removed when the patient was 15 years 8 months of age. A Hawley type retainer and fixed lingual retainer were then applied to the upper and lower dentitions, respectively. The removable and fixed retainers were removed at age 18 years 7 months.

Results achieved. Optimum intercuspation of teeth with parallel dental roots was achieved from the active orthodontic treatment (Figure 5). The skeletal Class I apical base relationship (Figures 4 and 6) was maintained. Acceptably good occlusion and facial profile were also maintained after the retention period of 2 years (Figures 1 and 6).

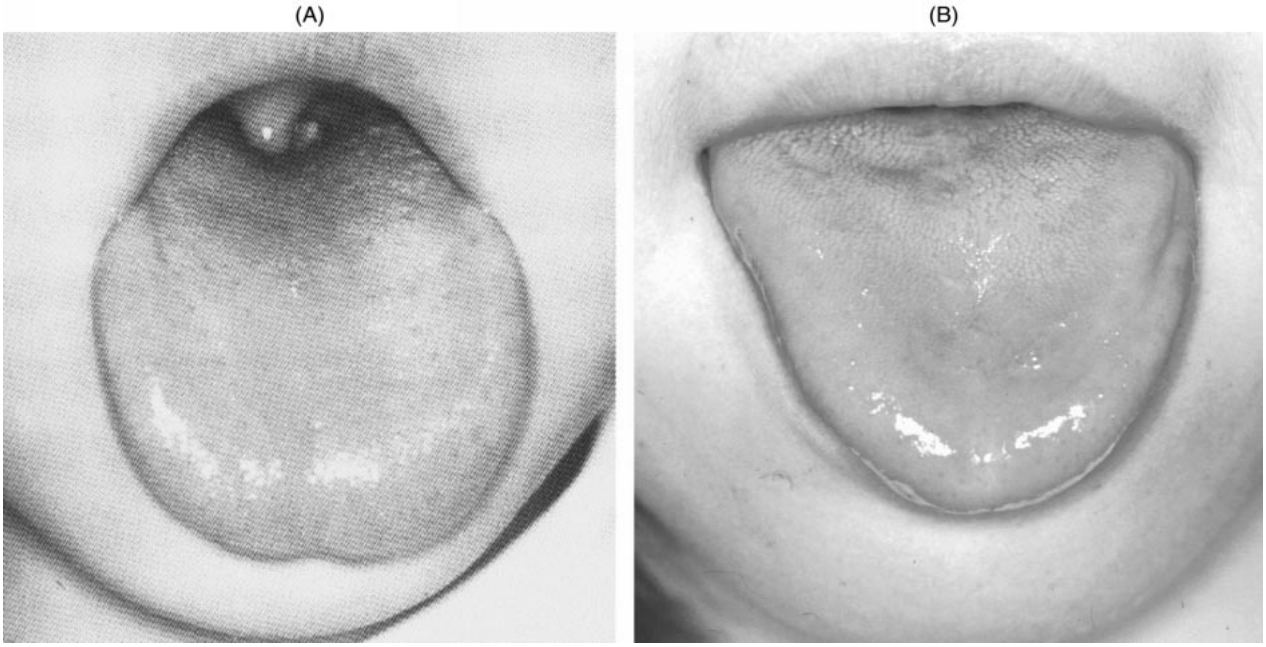

FIGURE 2. Photographs of protruded tongue. (A) Initial stage (6Y4M); (B) Just before edgewise treatment (11Y6M).

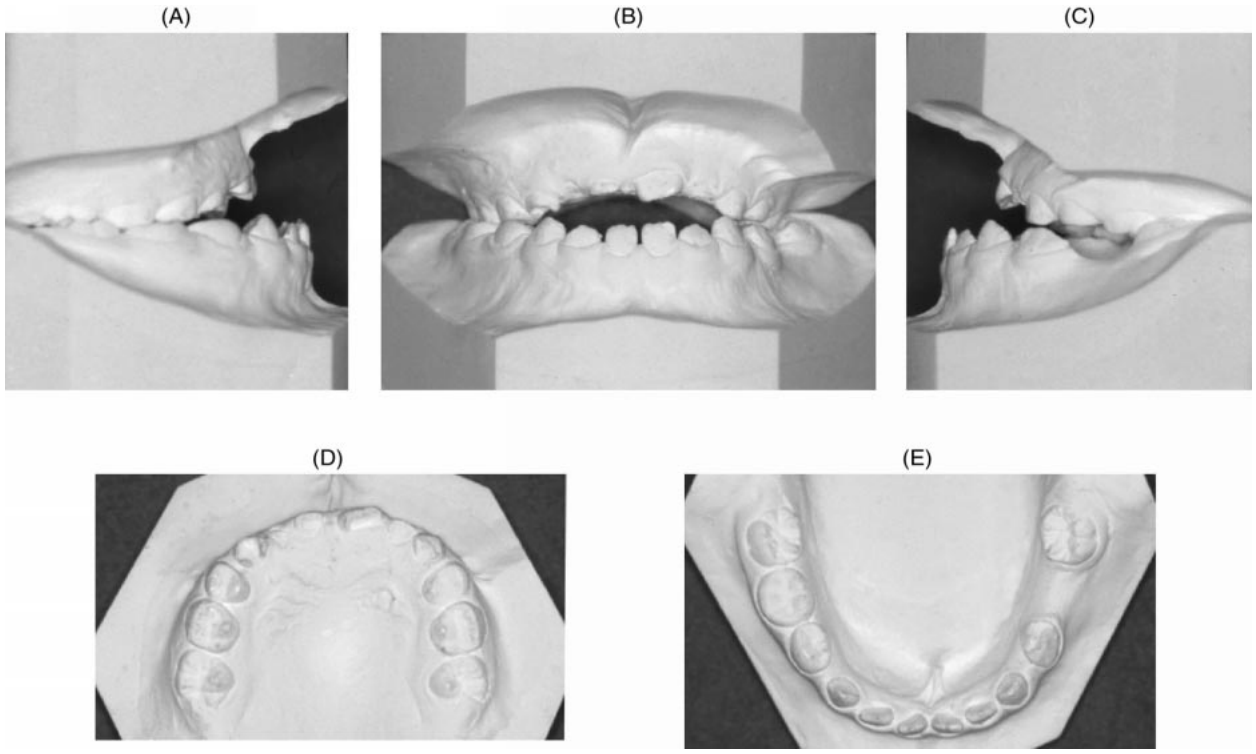

FIGURE 3. Photographs of dental casts at the pretreatment stage (6Y4M). (A) Lateral view on the right side; (B) Frontal view; (C) Lateral view on the left side; (D) Occlusal view of maxillary dentition; (E) Occlusal view of mandibular dentition.

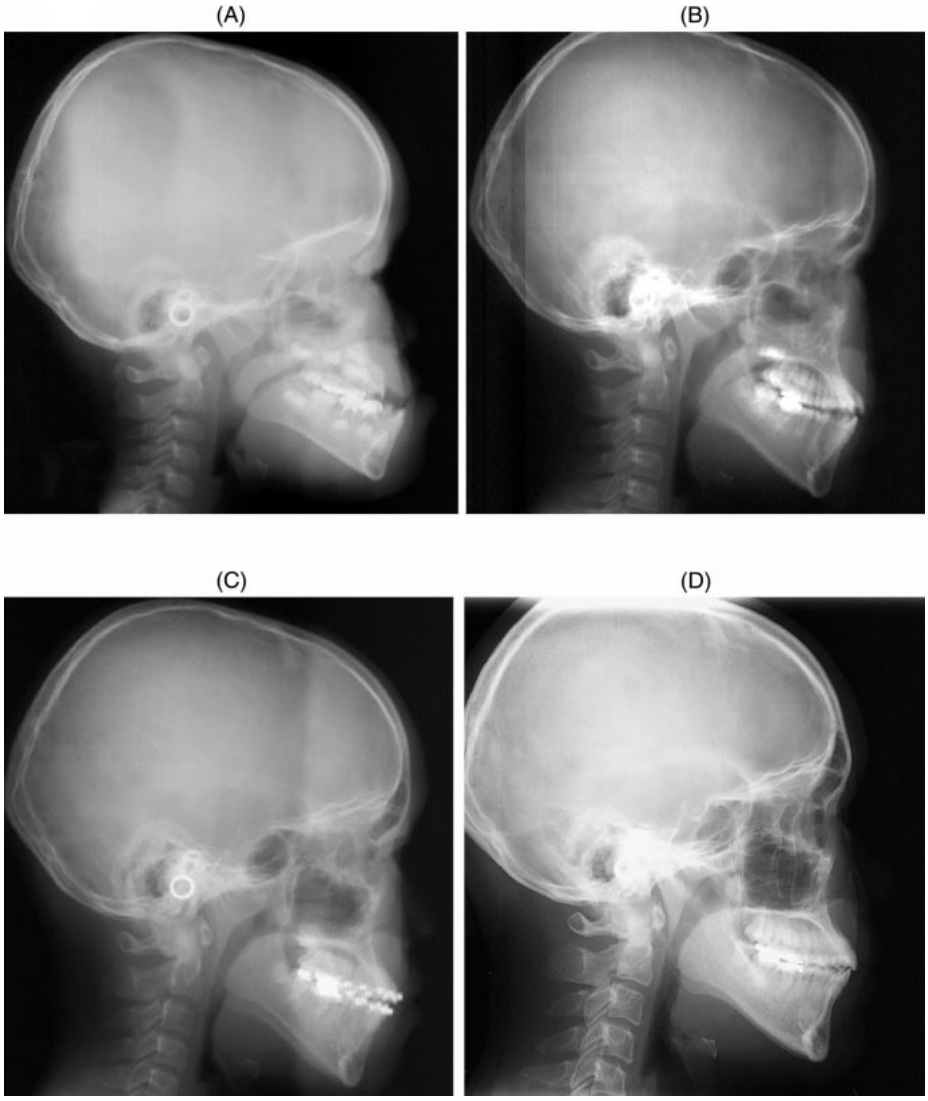

FIGURE 4. Cephalometric radiographs. (A) Initial stage (6Y4M); (B) Just before edgewise treatment (11Y6M); (C) Just before debonding (15Y6M); (D) After retention period (18Y7M).

TABLE 1. Cephalometric Analysis at the Initial Stage (6Y4M), Just Before Edgewise Treatment (11Y6M) and After the Retention Period (18Y7M)

	6Y4M		11Y6M		18Y7M	
Measurements	Value	Z-Score[a]	Value	Z-Score[a]	Value	Z-Score[a]
N-S (mm)	64.6	+0.8	67.9	+0.3	71.1	+0.9
Ar-Me (mm)	105.1	+4.9	106.9	+0.9	114.9	+1.4
N-Me (mm)	107.5	+0.7	120.5	−0.1	133.3	+1.5
SNA (degree)	83.8	+0.9	81.7	+0.3	80.3	+0.1
SNB (degree)	87.7	+4.3	82.1	+1.1	81.1	+0.7
MP-SN (degree)	34.9	−1.3	37.8	+0.2	38.7	+0.3
Gonial angle (degree)	137.5	+1.6	132.6	+2.2	130.7	+1.6
Overjet (mm)	−9.1	−8.9	0.9	−2.3	2.3	+0.7
Overbite (mm)	−6.0	−4.5	−0.9	−2.3	1.3	+1.1
U1-SN (degree)	108.0	+2.9	111.1	+0.7	109.1	+0.4
L1-MP (degree)	88.0	+0.1	84.6	−1.5	88.7	+0.7

[a] Z-score was calculated as (value − norm)/1 SD using norms and SDs of mean Japanese females according to the corresponding age.[10]

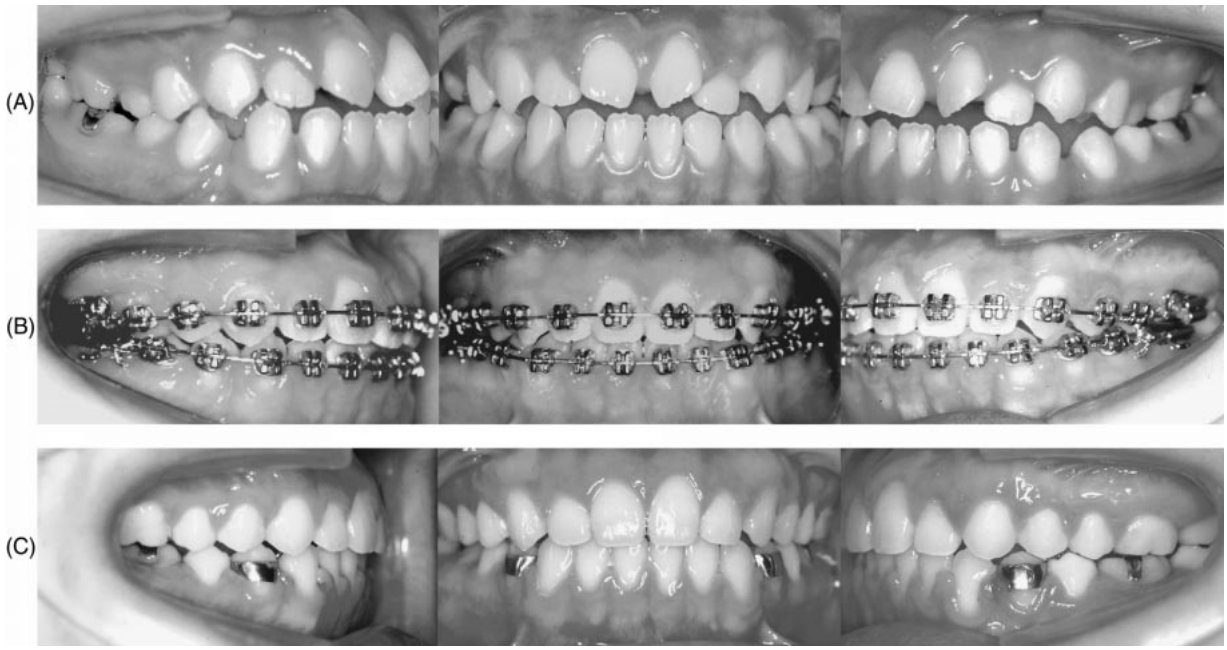

FIGURE 5. Intraoral photographs. (A) Just before edgewise treatment (11Y6M); (B) Just before debonding (15Y6M); (C) Just before removing of retainer (18Y6M); Left: Lateral view on the right side; Center: Frontal view; Right: Lateral view on the left side.

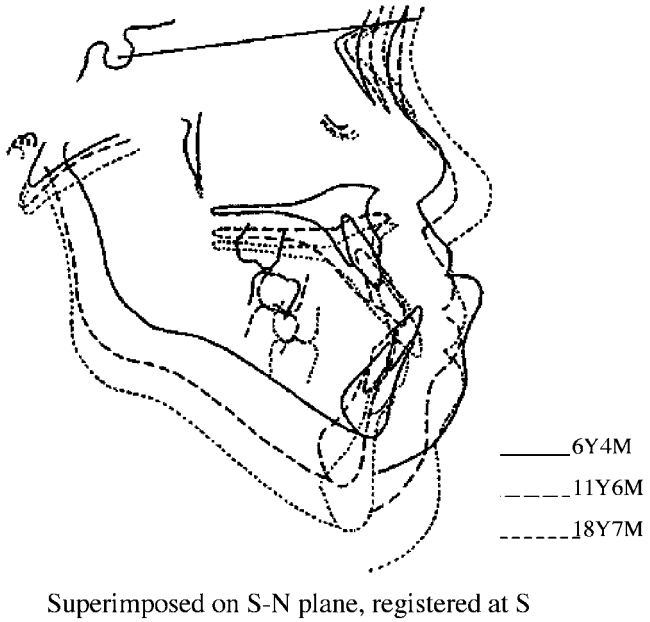

FIGURE 6. Changes from initial to post-retention stages on superimposed tracings of lateral cephalograms. Solid line: Initial stage (6Y4M); Dashed line: Just before edgewise treatment (11Y6M); Dotted line: After retention period (18Y7M).

DISCUSSION

A genetic diagnosis is now possible for Beckwith-Wiedemann syndrome.[12,13] A past radiographic cephalometric study, using patients who had and had not undergone tongue reduction surgery, showed that the anterior open bite and mandibular protrusion of patients who had undergone tongue reduction were improved.[8] The patients in this study showed similar improvements. The changes at the preadolescent period, following tongue reduction surgery and phase I orthodontic treatment using a chin cap and tongue crib, were dramatic as shown in Figures 4 and 6. Due to these effects, the patient's abnormal dentoskeletal pattern, including skeletal Class III and anterior open bite, became almost normal at the early stage of the adolescent period. As a result, phase II orthodontic treatment was successful, with the patient showing few signs of relapse 2 years after edgewise treatment.

Recently, a patient with Beckwith-Wiedemann syndrome who underwent surgical orthodontic treatment and was observed over a long period of time was reported in Japan.[14] Although the patient showed severe anterior open bite due to macroglossia, dentoskeletal disharmony was adjusted by a sagittal split ramus osteotomy without conducting tongue reduction. By 1 year after edgewise treatment, the patient showed a severe relapse with a 2 mm open bite despite the fact that acceptable occlusion was present at debonding.

Accordingly, tongue reduction conducted at a preadolescent period surgical orthodontic treatment may preclude the need for surgical orthodontic treatment, with virtually no relapse and resulting in stable occlusion. Both a previous study[8] and this case report suggest the efficacy of the tongue reduction in Beckwith-Wiedemann syndrome patients with macroglossia to improve the skeletal Class III and open bite. However, there is also a report showing that functional treatment using a pacifier and stimulation plate to the tongue during infancy resulted in no need for tongue reduction.[9]

Based on previous studies and our case report, we recommend that patients with Beckwith-Wiedemann syndrome undergo the following orthognathic treatment: (1) early functional treatment of the stomatognathic system; (2) if the effect is not adequate, tongue reduction should be carried out as early as possible during the early stage of the preadolescent period; and (3) orthodontic treatment, orthognathic surgery, or both, should be initiated as necessary.

ACKNOWLEDGMENTS

We extend our appreciation to Mr Virgil Hawkins of Osaka University for checking the English grammar of this paper.

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